

Fig 1

Fatal rupture of an oesophageal varix

THE PATHOLOGY AND MANAGEMENT OF PORTAL HYPERTENSION

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LONDON
EDWARD ARNOLD (PUBLISHERS) LTD

First published 1959

*Made and printed in Great Britain by
William Clowes and Sons, Limited, London and Beccles*

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INTRODUCTION

The upsurge of interest in vascular surgery which has occurred in the last twenty years has cast its mantle over the portal circulation Progress in the knowledge of this circulation has been hindered by its inaccessibility to the ordinary methods of examination and investigation used in the systemic blood vessels During the last few years the use of hepatic vein catheterisation, splenic puncture, portal venography and liver needle biopsy have contributed greatly to our knowledge of the portal circulation and its pathology It has not been possible to reproduce easily in animals the abnormal conditions found in man, so that much of the research into this circulation has had to be clinical research, carried out by physicians, surgeons and radiologists This volume is based on the experience of over two hundred patients seen during the last ten years, most of these patients have had surgical operations, and thus there has been an opportunity to study the pathology of the living, a study which is denied to many workers who only see their patients from the outside or when they have passed the terminal stages of their disease

Venous shunting operations for portal hypertension have now been in routine use for rather more than a decade, and their early technical problems have been overcome, there have been no major changes in the management of this condition during the last five years or so, and thus the time seemed appropriate to review the present position in the light of the experience which has been gained There are still many problems to be worked out both as regards the pathology and the treatment In the majority of cases little is known about the cause of the disease in the liver or the mechanical factors which lead to the hypertension, and the problem of the management of those patients in whom a shunt operation is either inadvisable or impossible remains as great as ever

It is the purpose of this book to serve as a guide to the clinicians who have to treat these cases, and at the same time to attempt to stimulate further investigation, both by experimental methods and by clinical experience, which will throw light on the many unsolved problems and thus in turn assist in the prevention and management of portal hypertension

During the last few years a voluminous literature of the subject has appeared but most of it records personal experience, often with very inadequate data both as regards numbers of patients and length of time for which they have been studied The clinician thus has great difficulty in assessing the present position amongst this maze of reports, and in this volume I have tried to set out briefly how the matter stands at the

moment, realising full well that great changes may take place in the future concerning the management of these patients. No attempt has been made here to review the whole series of my own patients, but a number of short case histories are recorded to illustrate special points.

I have not recapitulated the historical aspect of the subject as this has already been admirably reviewed by Child (1954). Two monographs describing series of cases have appeared during the last year by C.-A. Ekman (1957) giving the experience of 82 patients in the University Hospital at Lund, and by A. H. Hunt (1958) who recounts an experience of 250 patients at St. Bartholomew's Hospital, London. In addition the extensive clinical experience of Blakemore in New York and of Linton in Boston is described in a number of papers. Those who are interested in the experimental aspect of hepatic cirrhosis are referred to the comprehensive review by Kobak (1956).

My thanks are due to a great many people who have assisted in providing the experience on which this work is based. First of all come the patients, who have nearly all been most helpful in keeping in touch so that the follow up of their treatment has been almost complete. Next come the *general practitioners, physicians and surgeons who have referred their* patients and placed confidence in the team who are interested in this work at the Bristol Royal Infirmary. These are so numerous that it is impossible to mention them by name, but particular gratitude is due to a few physicians who have amongst them contributed a high proportion of the patients on whose study this work is based. To my assistants, anaesthetists and nursing staff I take this opportunity of expressing my deep debt of gratitude for their untiring help; any successful results of surgical treatment are due to their work more than to the actual technical steps of the operation; the radiologists and pathologists form indispensable members of the team, for without their co-operation very little clinical research is possible. Practically all the photographs in this book have [illegible] of the University [illegible] ity to [illegible] *ed by* courtesy of the editor of the *Journal of Physiology*.

It is a tragedy that patients who are otherwise fit still die of haemorrhage from oesophageal or gastric varices. An example of this is shown in the illustration opposite the title page. Many of these patients can be saved for years of active and useful life by timely surgical treatment, and if the work on which this monograph is based has saved the lives of a few of them, it will have at least some justification.

Bristol, 1958

REFERENCES

CHILD, C G (1954) *The Hepatic Circulation and Portal Hypertension* Philadelphia and London Saunders
'KMAN, C-A (1957) 'Portal Hypertension Diagnosis and Surgical Treatment" *Acta Chir scand*, Suppl 222, p 143
UNT, A H (1958) *A Contribution to the Study of Portal Hypertension* Edinburgh and London Livingstone P 230
KOBAK M W (1956) Experimental Surgery of Hepatic Cirrhosis" *Int Abstr Surg*, 102, 521-544

CHAPTER I

ANATOMY AND PHYSIOLOGY OF THE PORTAL SYSTEM

Gross anatomy

The portal venous system outside the liver consists of three main vessels and their tributaries The main vessels are the superior mesenteric vein and the splenic vein, which unite behind the neck of the pancreas to form the third main vessel, the portal vein After a course of 5 or 6 cm in an oblique direction upwards and to the right, this vein divides in the hilum of the liver into two main branches Occasionally a third branch comes off at the same level, an important point if the branches are being ligatured when making a portacaval anastomosis Venography has shown that the direction of the portal vein is much more oblique than is usually shown in anatomical text-books, and it may lie almost horizontal The distribution of the branches in the liver has been fully described by Couinaud (1953) and by Gans (1955) and affects the surgery of the liver rather than the extra-hepatic portal venous system

The tributaries of the main vessels are variable The two most important are the left gastric vein and the inferior mesenteric vein, for these vessels are the main channels by which there is an anastomosis with the systemic veins, draining as they do the two ends of the intra abdominal part of the alimentary canal

The left gastric vein is normally the largest vein which drains the stomach In about two-fifths of all people it enters the upper border of the splenic vein within 3 cm of the termination of the latter, in a similar proportion it joins the right border of the portal vein and in the remaining fifth it comes in just at the junction of the splenic and superior mesenteric veins The site of the termination of the inferior mesenteric vein bears similar proportions, in two-fifths into the splenic vein behind the pancreas, in two-fifths into the upper part of the superior mesenteric vein, and in the remaining fifth it joins the angle between these veins at their junction (Fig 2)

Portal-systemic anastomosis

From the point of view of pathology the anastomoses between the portal and systemic veins are important Normally these anastomoses are insignificant, but if there is obstruction to the flow of portal blood along its normal course they assume great importance

The stomach and oesophagus

The anastomoses at the cardiac end of the stomach which communicate with the oesophageal veins need particular attention Butler (1951) has made a very careful study of these communications He has shown that in the lower part of the oesophagus there are sub epithelial and sub mucous venous plexuses separated by the muscularis mucosae From the sub mucous plexus veins perforate the muscular coat and unite to form extrinsic veins and these perforating veins are provided with valves which direct the blood outwards The two plexuses in the oesophagus communicate

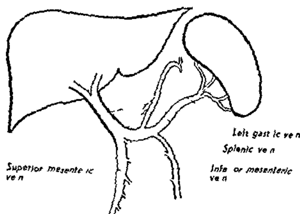


Fig 2 The main veins of the Portal System The common variations in the terminations of the left gastric and inferior mesenteric veins are shown by broken lines The boundary between the areas of distribution of the right and left branches of the portal vein in the liver corresponds to a line drawn vertically from the gall bladder fossa

freely with corresponding plexuses in the stomach and those communications which lie in the sub mucous layer may contain valves which direct the blood from the oesophagus to the stomach There are communications between the anterior and posterior tributaries of the left gastric vein and the corresponding veins which accompany the vagus nerves terminating in the azygos vein on the right side and the hemiazygos and left posterior bronchial veins on the left Most of the extrinsic veins join the azygos vein and its tributaries but a few drain into intercostal veins and communicate freely with the periaortic venous plexus Small oesophageal veins join the superior and inferior phrenic veins Butler found that India ink injected into the superior mesenteric vein of the foetus found its way into the sub epithelial plexus at the lower end of the oesophagus but

some of it also passed by the left gastric vein to the azygos and posterior bronchial veins

At the other end of the alimentary canal there are communications between the radicals of the inferior mesenteric vein in the sub-mucosa of the rectum and tributaries of both the middle and inferior rectal veins which drain into the systemic veins

The falciform ligament

Other important collateral channels occur in the falciform ligament Butler (1952) found large veins present in this site in two cases, in both a thick-walled branch came off the left branch of the portal vein and after running for some way in the falciform ligament it was continued as a dilated thin-walled vessel which drained into the veins of the anterior abdominal wall, and was in fact a greatly enlarged vein of Burow These veins, branches of the umbilical vein, are normally quite small, but are constant both in the foetus and in the adult Clinical observation confirms these [illegible]

wall some little way above the umbilicus Of the paired umbilical veins in the embryo, it is usually the left one alone that persists, so that normally the umbilical vein comes off the left branch of the portal vein, but occasionally, as was well shown in one of our venograms, it joins with the right branch of the portal vein

Elsewhere there are many small communications between the portal and systemic veins In the retroperitoneal tissues there are communications to the lumbar and to the azygos and hemiazygos veins, to the adrenal and renal veins and to the lower intercostal veins particularly by way of vessels in the lieno-renal fold of peritoneum In addition communications are to be found in the ligaments of the liver leading to the diaphragm, and draining mainly to the pericardio-phrenic veins

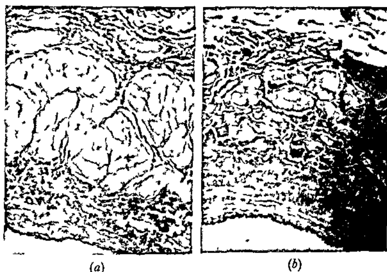
The portal vein itself is relatively thick-walled containing much more smooth muscle than any systemic vein of similar size (Fig 3)

Intra-hepatic circulation

In order to appreciate the pathology of some of the conditions which influence the portal circulation it is necessary to study the minute anatomy of the liver lobules, and much light has been thrown on this recently, particularly by the work of Knisely *et al* (1948) Branches of the hepatic *artery and the portal vein divide in the portal tracts* and enter separate sinusoids at the periphery of the liver lobule, but these sinuses unite and by the time the blood from these two sources reaches the parts of the sinusoids near the centre of the lobule it is intimately mixed The arterioles entering the lobules are much smaller than the corresponding

portal venules, and it is probable that the resistance in these arterioles leads to an equalisation of pressure in the sinusoids which are supplied by blood from both sources (Fig 4)

Injection studies have shown that in the portal tracts there are communications between branches of the hepatic arteries and the portal veins, their numbers vary in different animals but they probably become enlarged in cases of portal hypertension No definite evidence has been found of communications in the liver between the hepatic arteries and portal veins on the one hand and the hepatic veins on the other in the substance of the liver except through the sinusoids Where there is



(*a*) (*b*)

×60

severe fibrosis such communications probably form so that blood can flow through the liver without going through the sinusoids

In some animals the orifices of the hepatic veins as they enter the inferior vena cava are surrounded by smooth muscle fibres which exert a sphincteric action on the veins and may thus control the outflow of blood from the liver Such a mechanism has not been demonstrated in man

Physiology

The main function of the portal circulation is to convey some of the products of digestion from the alimentary canal to the liver *The liver*

takes up glucose and stores it as glycogen Some amino acids are carried to the liver and there undergo changes resulting in their detoxication before they are released into the systemic circulation. There is very little factual knowledge of the control of this circulation The inflow is dependent on the flow of blood in the splanchnic branches of the aorta, which in its turn is under the control of the sympathetic nervous system It is possible that there is also some control of its outflow from the liver Daniel and Prichard (1951) have shown by radiographic means that in

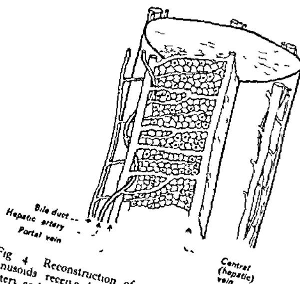


Fig 4 Reconstruction of a liver lobule The sinusoids receive blood from both the hepatic artery and the portal vein, in the portal tracts the arteries are not nearly so straight as the veins

rats there are considerable variations in the blood flow through the liver Sometimes the portal blood flow is confined to short wide sinusoids situated centrally in the liver, at other times the flow is evenly distributed right out to the periphery of the organ and the rate of flow is much slower The significance of these changes is not known and they require further investigation (Fig 5)

The pressure gradient between the portal and hepatic veins is small, in health not more than 10 mm Hg, so that very little obstruction to the flow through the liver will tend to bring about a rise in the portal venous pressure.

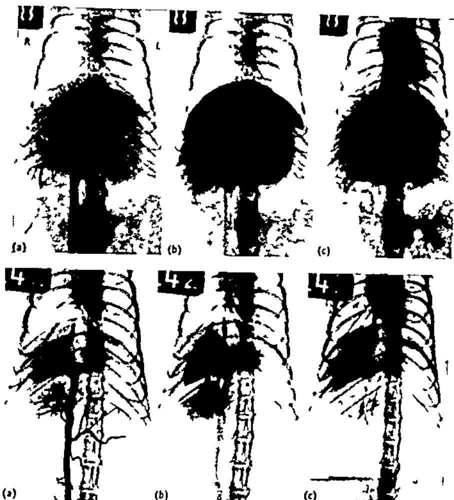


Fig 5 The portal blood flow in the rat Studies made by injection of an opaque medium into the portal circulation In the upper series all parts of the liver were filled In the lower series only the central area of the liver was filled and the blood passed through more quickly (Daniel R M and Pritchard M M L)

Experimental portal hypertension

Attempts to produce portal hypertension in animals have in the main been unsuccessful Sudden occlusion of the portal vein is rapidly fatal in cats dogs and rabbits but if a venous anastomosis is made to prevent portal congestion recovery ensues However, the macaca monkey will survive portal vein ligation but the portal pressure falls to normal within six to ten days (Child *et al*, 1950) Slow occlusion of the vein may be obtained

by wrapping cellophane around it but in animals the collateral circulation which forms is adequate to maintain the portal pressure within normal limits

Measurement of portal pressure

In man it is no simple matter to measure the portal venous pressure and in fact it was only just over twenty years ago (Rousselot 1936) that it was first recorded It can only be done either at a laparotomy or by indirect means

At a laparotomy the simplest method is the use of a manometer filled with saline connected by a three way tap to a needle or canula inserted into one of the veins of the portal system If one of the smaller veins is used a needle is liable to damage the wall and more reliable readings are obtained with the use of a small canula This can be placed in the right gastro epiploic vein or in a vein of the mesentery of the small intestine or the great omentum which may be ligatured afterwards When a recorder with a pressure head of the transducer or strain gauge type is available tracings can be obtained with a needle in either the portal or splenic vein The fact that the abdomen is open when the measurements are made probably does not affect the reading appreciably but normally intra abdominal pressure is transmitted to the veins and will thus influence the portal venous pressure When making a reading it is important to ensure that nothing is obstructing the vein between the point of reading and the liver and there should be a definite excursion of the fluid corresponding with respiratory movement (Fig 6)



Fig 6 A saline manometer used for recording the portal venous pressure

Intra-splenic pressure

Atkinson and Sherlock (1954) have shown that the intra splenic pressure is a reasonably good indication of the portal venous pressure A needle

is inserted through the parietes into the spleen and, when blood drips from the needle, it is connected to a manometer The measurement can conveniently be made at the same time as a trans-splenic portal venogram These writers found levels ranging from 3 to 17 mm Hg in 14 patients without evidence of portal hypertension, but in cases of portal hypertension the range was from 17 to 35 mm Hg The intra splenic pressure exceeded the portal venous pressure estimated by hepatic vein catheterisation by 2 to 6 mm Hg

Hepatic vein pressure



Fig 7 A cardiac catheter wedged in a tributary of a hepatic vein in order to record the portal venous pressure

Another indirect means of measurement of the portal pressure is by means of hepatic vein catheterisation and wedging a catheter with a terminal opening in a tributary of a hepatic vein (Fig 7) When this is done it is assumed, in cases of intra hepatic obstruction, that the pressure immediately proximal to the catheter will build up to the portal pressure In cases of extra hepatic obstruction this build up of pressure will not occur, so that as a means of measuring the pressure this is only useful when there is disease of the liver without extra-hepatic obstruction This method is occasionally of value in distinguishing between intra and extra-hepatic obstruction but otherwise has little place in routine clinical practice, if hepatic vein catheterisation is being done as a means of measuring the liver blood flow, a pressure recording may be made at the same time In 24 patients without liver disease a mean pressure of 6 8 mm Hg was found but in 11 patients with cirrhosis of the liver, levels varied from 15 6 to 35 2 mm Hg with a mean of 22 0 mm Hg (Paton *et al*, 1953)

Allison (1951) made measurements of the pressure in oesophageal varices by direct puncture using a needle through an oesophagoscope The method is difficult in practice, and in any case the pressure in

these vessels is not a true indication of the pressure in the veins nearer the liver.

Normal portal pressure

A number of measurements of portal venous pressure has been made by means of a saline manometer during operations on patients whose livers and portal systems were to all appearances quite normal In our series the range was from 7 5 to 12 mm Hg taking the level of the portal vein as zero Other workers have found rather wider limits Gray (1951) taking the level of the body of the adjacent vertebra as the base line found figures varying from 10 to 16 mm Hg If the portal vein is temporarily occluded during a laparotomy, the pressure rises rapidly to something between 40 and 50 mm Hg and remains at this level while the obstruction is maintained

Liver blood flow

Bradley and his colleagues (1945) devised a means of measuring the liver blood flow in man by the bromsulphalein excretion method, this depends on the absorption of this substance by the liver alone Intravenous infusion maintains a constant level in the systemic circulation and samples taken from a catheter in a hepatic vein indicate the amount removed by the liver and from this the flow can be calculated More recently, attempts to measure the flow by the use of radio-active isotopes have been made (Riddell *et al*, 1957) Normally the flow is about 1500 ml per min in adults These investigations record the total liver blood flow, and a satisfactory method has yet to be devised by which the relative hepatic artery and portal vein flows can be determined in man

REFERENCES

ALLISON, P (1951) "The Measurement of Blood Pressure in Oesophageal Varices" *Thorax*, 6, 325–327

ATKINSON, M and SHERLOCK, S (1954) "Intrasplenic Pressure as Index of Portal Venous Pressure" *Lancet*, 1, 1325–1327

BRADLEY, S E, INGELFINGER, F J, BRADLEY, G P and CURRY, J J (1945) "The Estimation of Hepatic Blood Flow in Man" *J clin Invest*, 24, 890–897

BUTLER, H (1951) "The Veins of the Oesophagus" *Thorax*, 6, 276–296

—— (1952) "Gastro-oesophageal Haemorrhage in Hepatic Cirrhosis" *Thorax*, 7, 159–166

CHILD, C G, MILNES, R F, HOLSWADE, G R and GORE, A L (1950) "Sudden and Complete Occlusion of the Portal Vein in the Macaca Mulatta Monkey" *Ann Surg*, 132, 475–495

COUINAUD, C (1953) "Étude de la Veine Porte intra-hépatique" *Pr méd*, 61, 1434–1438

DANIEL, P M and PRICHARD, M M L (1951) "Variations in the Circulation of the Portal Venous Blood within the Liver" *J Physiol*, 114, 521–537.

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REFERENCES

ALLISON, P (1951) "The Measurement of Blood Pressure in Oesophageal Varices" *Thorax*, 6, 325–327

ATKINSON, M and SHERLOCK, S (1954) "Intrasplenic Pressure as Index of Portal Venous Pressure" *Lancet*, 1, 1325–1327

BRADLEY, S E, INGELFINGER, F J, BRADLEY, G P and CURRY, J J (1945) "The Estimation of Hepatic Blood Flow in Man" *J clin. Invest*, 24, 890–897

BUTLER, H (1951) "The Veins of the Oesophagus" *Thorax*, 6, 276–296

—— (1952) "Gastro-oesophageal Haemorrhage in Hepatic Cirrhosis" *Thorax*, 7, 159–166

CHILD, C G, MILNES, R F, HOLSWADE, G R and GORE, A L (1950) "Sudden and Complete Occlusion of the Portal Vein in the Macaca Mulatta Monkey" *Ann Surg*, 132, 475–495

COUINAUD, C (1953) "Étude de la Veine Porte intra-hépatique" *Pr méd*, 61, 1434–1438

DANIEL, P M and PRICHARD, M M L (1951) "Variations in the Circulation of the Portal Venous Blood within the Liver" *J Physiol*, 114, 521–537

Gans, H (1955) "*Introduction to Hepatic Surgery*" Elsevier Publishing Company Amsterdam

Circula

Danske Videnskab Selskab Biol Skrifter IV, nr 7

Paton, A, Reynolds, T B and Sherlock, S (1953) "Assessment of Portal Venous Hypertension by Catheterisation of Hepatic Vein" *Lancet*, **1**, 918–921

Riddell, A G, Griffiths, D B, McAlister, J M and Osborn, S B (1957) "The Measurement of Liver Blood Flow with Colloidal Radiogold (^{198}Au)" *Clin Sci*, **16**, 315–324

Rousselot, L M (1936) "The Role of Congestion (Portal Hypertension) in So called Banti's Syndrome" *J Amer med Ass*, **107**, 1788–1793

CHAPTER II

PATHOLOGY OF PORTAL HYPERTENSION

The pressure in the portal circulation may rise either as a result of increased inflow through the splanchnic vessels, or of increased resistance to the outflow There has been some discussion regarding the significance of increased inflow, if the outflow is capable of carrying an extra volume of blood the additional inflow is unlikely to influence the pressure When the resistance to the inflow is diminished, as for example in the case of an arterio-venous fistula in the splanchnic circulation then the portal pressure will inevitably rise Evidence has been produced (Leather, 1957) that in cases of gross splenomegaly without obstruction to the portal blood flow there may be increased portal pressure, but this is based mainly on measurements of intra splenic pressure, and in such cases this may not give a true indication of the portal venous pressure

Increased resistance to the outflow of blood from the portal system is caused by lesions affecting the portal vein (pre- or extra hepatic obstruction), lesions of the liver (intra-hepatic obstruction) or lesions which raise the pressure in the hepatic veins (post-hepatic obstruction) The last group includes thrombosis of the inferior vena cava, tricuspid valve stenosis or incompetence, and constrictive pericarditis Such back pressure leads to irreversible changes in the liver which add an intra-hepatic obstruction to the post-hepatic cause, and care has to be taken in distinguishing between the two or the mistake may be made of treating the hepatic lesion without realising that it is not the primary cause of the portal hypertension In these post-hepatic lesions the portal pressure is not greatly raised, the spleen is rarely palpable far below the costal margin, and a collateral circulation is not much in evidence In other words, the portal hypertension plays a minor role and as such does not require special treatment

There are a few patients in whom there is portal hypertension, who develop a collateral circulation, but in whom no evidence of either increased inflow or obstruction to the outflow can be found It is possible that there may be a functional obstruction to the outflow, in the form of spasm of the portal vein or its branches in the liver The view has also been expressed that in some cases there may be obstruction to systemic veins which normally have anastomoses with the portal circulation, e g the azygos vein, and this leads to the diversion of blood by way of these anastomoses into the portal circulation, it seems unlikely that such a mechanism is ever significant

In this monograph, portal hypertension is taken to be a rise in the pressure throughout the portal venous system. However, it occasionally happens that in cases of obstruction to the splenic vein, the hypertension is confined to the veins of the spleen and the upper part of the greater curvature of the stomach. These cases should be called "splenic hypertension". Though they are rare, their recognition is important for they can be cured by removal of the spleen, haematemesis is often the presenting symptom, they may have oesophageal varices, but the collateral circulation tends to cross the stomach and drain into the left gastric vein if that vessel enters the main veins beyond the obstruction. The condition can only be diagnosed accurately by venography, both by injection into the spleen and into a tributary of the superior mesenteric vein, which will show the limits of the obstruction.

Extra-hepatic obstruction

Extra-hepatic obstruction of the portal blood flow may be primary or secondary, and may be due to thrombosis in the lumen of the portal vein or to pressure or invasion from without. When it is due to pressure or invasion from outside the vein, the treatment is that of the cause and the portal hypertension does not require special attention. As an example, patients with malignant lymph nodes in the portal fissure may present with splenomegaly. Similarly, malignant tumours of the stomach or pancreas may obstruct the portal or splenic veins and cause a general portal hypertension or a localised splenic hypertension.

Apart from the conditions which carry an increased risk of intravascular thrombosis, the following causes of portal vein thrombosis are recognised:

1. Congenital
2. Injury
3. Secondary to intra hepatic obstruction
4. Portal pylephlebitis caused by inflammation in the organs drained by the portal system

1 *Congenital*. The majority of patients with extra-hepatic obstruction and normal liver parenchyma are children or adolescents. For this reason it has been suggested that the condition is a true congenital abnormality, with failure of proper development of the portal vein. This may well be so in rare instances, but the observations of Gibson and Richards (1955) and of Parker and Seal (1955) suggest that much more frequently the cause is an intra-vascular thrombosis of the portal vein. It is suggested that this is particularly likely to occur at or soon after birth, either as an extension of infection from the umbilicus, or that the normal process of obliteration of the umbilical vein and ductus venosus extends into the portal vein. A few cases are on record in which a definite history of

umbilical sepsis has been obtained, and in the following case, in which an acute suppurative arthritis of the hip occurred during the first week of life, the source of infection was probably the umbilicus

S S, female, had within a week of birth, abscesses over her left buttock, left ankle, and on her head, these healed but at the age of 20 months, when she first attempted walking, it was found that she had a pathological dislocation of the left hip, which was treated by traction and a caliper, and later by an osteotomy When just 2 years old she had her first haematemesis, and

in the oesophagus above the line of section

Occasionally, the obstruction in the portal vein is represented by a short stricture, which may well be a true congenital abnormality

removed and an orthodox end-to-side splenorenal anastomosis performed During the five years since then he has remained quite well, but, at the age of 22, some varices are still present in his oesophagus

2 *Injury* Injury of the portal vein causing obstruction by scarring or by thrombosis is rare, but cases are on record in which it has been injured during a cholecystectomy and the patients have subsequently developed portal hypertension Ligature of the portal vein, however, does not always give rise to manifestations of portal hypertension, and

3 *Secondary to intra hepatic obstruction* In patients with long-standing

the blood in the portal vein may become almost stationary Both these factors predispose to thrombosis As long as this thrombosis is confined



Fig 8 Venogram by intra splenic injection showing a short stricture of the portal vein



Fig 9 Venogram by intra splenic injection The portal vein is narrowed by thrombosis In addition the left branch of the portal vein was obliterated by thrombosis A large left gastric vein leads to a mass of varices around the cardia

to the portal vein it may pass unnoticed clinically provided that the patient has developed an adequate collateral circulation, but if the process extends into and completely obstructs the superior mesenteric vein, infarction and gangrene of the small intestine is an almost inevitable sequel Many patients who die of hepatic cirrhosis are found to have some degree of thrombosis in the portal system Hunt and Whittard (1954) have drawn attention to the importance of this complication (Fig 9)

In a series of 80 patients with intra-hepatic obstruction whose portal veins have been explored and a portacaval anastomosis performed, mural thrombi partly occluding the lumen have been found nine times, in spite of the fact that in most cases the presence of a lumen had been demonstrated by preliminary venography Splenectomy appears to be a factor predisposing to thrombosis in the portal vein In all 3 patients in this group who had had a previous splenectomy, mural thrombi were present in the portal vein and in 1 the lumen was divided into two almost equal halves When the lumen becomes completely obliterated the risk of haemorrhage or portal systemic neuropathy is increased Such a state of affairs renders a portacaval anastomosis impossible, and the risk of this complication is an impressive argument against delaying this operation if the patient has had a severe haemorrhage and his liver function is good Before preliminary routine venography in our series the portal vein was explored in two cases of intra-hepatic obstruction and was found to be completely occluded

J O , male, age 29, had had his spleen removed following three haematemeses four years previously Intermittent bleeding persisted, and on exploring the region of the portal vein it was found to be completely thrombosed, and the pressure in a jejunal vein was 32 mm Hg No venous anastomosis was possible so an oesophageal transection was performed, apart from one slight haemorrhage he has remained well for the succeeding three years

A W , male, vomited blood for the first time at the age of 16, and shortly after this his spleen was removed He had no further haemorrhage for ten [illegible]

operation on his varices

These cases demonstrate not only the fact that patients may show no symptomatic evidence of thrombosis of the portal vein but the uselessness of splenectomy in treating this condition They also indicate that a short-term follow-up is quite valueless

4 *Portal phlebitis* Before the introduction of modern chemotherapeutic and antibiotic agents suppurative portal phlebitis was almost *invariably fatal* The source of the infection is most frequently the appendix, but in a number of patients its origin cannot be located (Shaldon, 1958) as in the following example Nowadays, as a result of treatment, the original infection may be overcome, but it will leave the patient with thrombosis of the portal vein This may be only a mural thrombosis or it may fill the lumen completely, *after a matter of months or years the* manifestations of portal hypertension will become evident



Fig 10 Venogram by intra splenic injection There has been thrombosis of the portal and splenic veins following suppurative portal phlebitis

W A, male, age 45, had fourteen months before *his first haematemesis*, a serious febrile illness which lasted for ten weeks and started with lower right-*sided abdominal pain His appendix had been removed fourteen years* previously On the thirteenth day of his illness, rigors commenced and on the twenty-second day he had jaundice which lasted four days A laparotomy on the twenty-sixth day revealed turbid intra peritoneal fluid but no source of his infection

Following the haematemesis his spleen was just palpable, he had gastric and oesophageal varices, and venography showed thrombosis of both splenic and portal veins (Fig 10) While awaiting operation he had another severe haemorrhage At operation the liver was normal, adhesions were present round the portal vein, and the pressure in a jejunal vein was 30 *mm Hg* The spleen was removed and a gastric transection performed The follow-*up is only of six months' duration*, but so far he has remained well

Thrombosis of the portal vein leads to great enlargement of small vessels which are normally insignificant, but which by-pass the obstruction, sometimes a degree of recanalisation of the vein takes place The result is a meshwork of irregular vessels, of varying size, but sometimes there are larger vascular spaces, giving the region much the appearance of a venous angioma, and the condition has been called "cavernous transformation of the portal vein" The extent of this change will be determined by the extent of the original thrombosis It may be confined to the extra-hepatic part of the portal vein, or it may be carried along many or all of the branches of the vein in the portal tracts of the liver (Fig 11) If

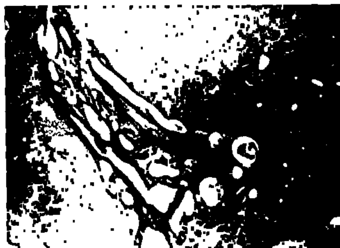


Fig 11 A portal tract in the liver of a girl who died of haemorrhage at the age of six, there has been extensive development of collateral venous channels forming the so-called cavernous transformation The liver parenchyma is normal × 30

the thrombosis also affects the splenic vein such a cavernous mass will be found extending along the upper border of the pancreas

The umbilical vein is normally a branch of the left branch of the portal vein in the liver, when a collateral circulation develops in the falciform ligament, and dilated veins appear in the subcutaneous tissues around the umbilicus, it is an indication that the obstruction is intra-hepatic In cases of thrombosis of the portal vein in its extra hepatic part, the obstruction is proximal to the junction of the umbilical vein and the left branch of the portal vein, and thus in this type of obstruction a "Caput Medusae" *will not form*

There must be considerable resistance to the flow of blood through the

tortuous channels in cases of cavernous transformation, for, although some opaque solution when injected into the portal circulation reaches the liver the portal pressure is often astonishingly high, frequently being above 30 mm Hg In children with this condition the liver has certai characteristics which can be recognised, it is particularly soft in consistency and has a brick red colour rather than the normal plum colour, presumably this is due to the fact that a much higher proportion than normal of its blood supply is arterial, these characteristics have not been seen in adults

There still seems to be some doubt whether deprivation of an otherwise normal liver of its portal venous blood supply is detrimental to its function or leads to permanent changes in its parenchyma As a result of the ex perience of 25 cases of primary extra hepatic obstruction in which liver biopsies and biochemical investigations are available, I am very doubtful whether this deprivation of venous blood leads to any disease in the liver itself Some of these patients have been studied for many years and there has been no evidence of progressive liver damage (see Chapter VI) In fact the only symptom from which these patients suffer apart from blood changes due to hypersplenism is haemorrhage from the varices of their collateral circulation Most of them if untreated will eventually d- as a result of haemorrhage Satisfactory treatment is no easy matter bu if the danger of haemorrhage can be overcome, these patients will lea normal active lives

Intra-hepatic obstruction

Many pathological conditions of the liver cause obstruction of the blood flow through it Owing to the high pressure gradient between the hepatic artery and the hepatic veins this obstruction needs to be severe before there is any appreciable reduction in the arterial blood flow through the liver and this obstruction does not influence the general arterial pressure By contrast the low pressure gradient between the portal and hepa veins results in the flow being very easily obstructed, and as the port circulation is a closed circulation except between capillaries, this obstruc tion leads at an early stage to a rise in pressure in the portal circulation

The conditions in the liver which cause obstruction to the portal blood flow may be temporary or permanent, it is probable that any condition which causes tension inside the liver capsule will hinder the flow of portal blood Thus acute hepatitis or fatty infiltration may lead to a transient rise in portal blood pressure which will subside as the liver returns to normal In 40 patients who had acute hepatitis Reichman and Davis (1957) recorded the intrasplenic pressure during the acute illness and again during convalescence and found that in all except 6 cases there was a sig nificant fall in the convalescent period varying from 14 to 64 per cent I 5 cases however a higher pressure was found during convalescence thar during the acute phase of the illness Conditions which set up a fibrous

response in the liver give rise to a more permanent portal hypertension, but it has been found that in cases where the liver is extensively replaced by fibrous tissue the portal pressure may be within the normal range or only slightly elevated The reasons for this will be discussed later

There are many causes of connective tissue proliferation in the liver which leads to fibrosis In some there has been necrosis of liver cells with collapse of the connective tissue framework, and proliferation of the surviving liver cells, often in irregular nodules The importance of fatty deposits in the liver cells is not clear In many cases of portal hypertension with hepatic fibrosis it is quite impossible to lay the blame on any particular etiological factor

Any of the following may be precursors of portal hypertension

1 *Virus hepatitis* Most cases of this infection clear up completely and it is not known why a few progress to cirrhosis, but nutritional factors may play a part (Himsworth and Glynn, 1944) Dible (1951) has followed the changes by means of serial liver biopsies It is probable that many cases are due to this cause, but the acute illness is not recognised as hepatitis because evident jaundice has been absent Symptoms of portal hypertension appear between two and ten years after the acute attack

M E , female, had an acute illness at the age of 33 lasting nine weeks, with jaundice Nine years later she attended hospital on account of pain in the left loin, and was found to be so anaemic that a blood transfusion of 3 pints was administered Three years later, at the age of 45, she had her first haematemesis which required a transfusion of 3 pints of blood, but her next attack eight months later necessitated the administration of 8 pints of blood Her liver function tests were good, intra splenic pressure was 25 mm Hg and venography showed a patent portal vein When an end to side porta caval anastomosis was made, the pressure in the portal vein before making the shunt was 29 mm Hg

2 *Nutritional deficiency* There is much experimental evidence that a low protein diet causes diffuse hepatic necrosis and post necrotic scarring This may occur in spite of an adequate calorie intake In many countries, particularly in Africa and the East, poor nutrition has been blamed for the high incidence of cirrhosis of the liver and portal hypertension but it may be that the poor nutrition increases the liability to virus infection In Southern Rhodesia, hepatic cirrhosis is much the most common cause of haematemesis (Gelfand, 1957) It is possible that the cirrhosis associated with ulcerative colitis or with fibro-cystic disease of the pancreas may be due to deficiency of protein

3 *Alcoholic cirrhosis* It is not definitely established whether there is a direct toxic effect, or the liver changes are due to a nutritional deficiency in those who take much alcohol This is, however, an important factor in a proportion of cases, and adversely affects the prognosis after a shunt operation if they continue their alcohol consumption

H D, male, age 39, had for the previous twenty years been in the habit of taking 4 pints of beer and 5 pints of cider a day. During the last eight years he had five attacks of haematemesis. At operation his liver was normal in size but hard with a nodular surface, and his portal pressure was 27 mm Hg. After a portacaval anastomosis, his spleen which had been enlarged to three fingerbreadths below the costal margin, became impalpable. However, he declined to reduce his alcohol consumption below 2 pints each of beer and cider, and died of liver failure two and a half years later.

4 *Haemochromatosis*. In this condition the accumulation of iron in the liver leads to extensive fibrosis, and thus to portal hypertension.

M F, male. This patient was boarded out of the Army on account of haemochromatosis in 1940 at the age of 32. In 1949 there were two attacks of severe haematemesis. At a laparotomy the liver was cirrhotic, the spleen moderately enlarged, and the pancreas brick-red in colour. At that time there was no glycosuria. The splenic artery was tied and an omentopexy performed. Two and a half years later he was found to have diabetes. In 1954 he had another haemorrhage and ascites appeared, and following further haematemesis he died five and a half years after his operation. At post mortem there was multilobular cirrhosis with multiple nodules of primary carcinoma of the liver.

5 *Hepato-lenticular degeneration*. Here there is an excess of copper deposited in the liver, and evidence of portal hypertension may be found.

C L, female, first noticed clumsiness of her hands at the age of 15, and soon after this found that she could not pronounce words clearly. The following year the diagnosis of hepato lenticular degeneration was made, the cornea showed typical Kayser-Fleischer rings and her serum copper was found to be 47 mg/100 ml. A year later she had her first haematemesis. Investigation showed no gross disturbance of liver function, large varices and a patent portal vein. While in hospital further bleeding occurred and she received 21 pints of blood by transfusion in six days. She was then transferred to the Bristol Royal Infirmary and two days later an end-to side portacaval anastomosis was performed. Her portal venous pressure was 43 mm Hg. During the year since her operation no further bleeding has occurred.

6 *Bilharzia*. The liver is involved by infection by *Schistosoma mansoni* and *Schistosoma japonicum*. The ova obstruct the branches of the portal vein and give rise to a periportal fibrosis. This is one of the common causes of portal hypertension in South America.

A S, male, was born in 1938 in Poland and was taken to Russia in 1941. He reached Persia in 1942, when he suffered from dysentery, in 1943 he was in India and then spent the years from 1943 to 1950 in Rhodesia and Tanganyika, where he had malaria. He reached England in 1950 at the age of 12 years and was complaining of lassitude and had persistent fever. A laparotomy showed cirrhosis of the liver due to Schistosomiasis mansoni. In 1952 he had a severe haematemesis for which transfusion was required. His liver was moderately and his spleen was grossly enlarged. Fine mottling was shown radiologically in both lungs. After a course of antimony tartrate, a portacaval anastomosis was performed. The portal pressure was 16 mm Hg and fell to 11 mm Hg after the anastomosis. The lymph nodes round the

portal vein were much enlarged Histology showed lesions containing large giant cells in the lung, liver and a lymph node, in the liver was also portal infiltration by lymphocytes and plasma cells, but no marked fibrosis A few degenerate parasites were demonstrated in the liver (Figs 12a) During the five years since his operation he has remained very well, and his liver and spleen have ceased to be palpable

7 *Biliary cirrhosis* Chronic biliary obstruction due, for example, to gall stones, or injury to the common bile duct, leads to cirrhosis of the liver which in turn causes portal hypertension If the obstruction is in the extra-hepatic passages, surgery should be undertaken for its relief, and operative treatment for the hypertension must be required only very rarely

8 *Congenital absence of the bile ducts* A particular form of biliary cirrhosis is due to congenital absence of the bile ducts Most of these patients die during the early months of life, but a few survive the initial obstructive jaundice and it appears that the alimentary canal takes on some of the excretory functions of the liver, however, these patients early develop the typical brown complexion of the cirrhotic, and ascites They also get oesophageal varices and may have haematemesis The liver function is too poor for any surgical treatment to be considered and they usually die of liver failure

Congenital fibrosis of the liver

There is a rare form of obstruction which occurs in children (Park 1956) It is of the pre-sinusoidal type, the portal tracts are large and contain not only an excess of fibrous tissue, but very many small bile ducts, some of which may penetrate, without any sheath, into the liver parenchyma and often contain inspissated bile The liver is enlarged and firm and when exposed at operation has a peculiar appearance, its surface is quite smooth, but it is mottled, many small areas of purple liver parenchyma being separated by a grey or yellow network which represents the fibrous tissue These children have no symptoms except haematemesis, and the changes in the liver do not appear to be progressive The portal pressure is high, but they do well after a portacaval anastomosis Four such patients have occurred in this series, 3 girls who had their first haemorrhages at the ages of 3 and 5 and 17 respectively, and a boy who bled for the first time at the age of 6 One of these is described in greater detail

L S, female, was found to have an enlarged liver at the age of 2, when admitted to hospital for an umbilical hernia Haematemesis occurred when she was 3, recurred twice when she was 9 and again at the age of 11 The liver was a little enlarged and firm, the spleen extended three fingerbreadths below the costal margin, but all liver function tests were normal At the age of 12 a portacaval anastomosis was performed, her portal vein pressure was 24 mm Hg The liver appearance both macro and microscopically was typical as described above (Fig 12b)

Fig 12 Gross and microscopic appearances of the liver in some conditions causing intra hepatic obstruction Photographs and biopsies taken at operation Histology—Mallory stain, ×30

(*a*) Patient A S, age 14 Schistosomiasis The liver is enlarged, dark-coloured and firm The granuloma contains a few giant cells Portal pressure 16 mm Hg

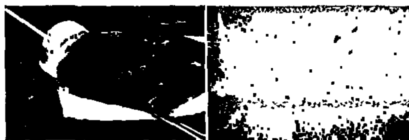
(*b*) Patient L S, age 12 Congenital fibrosis of the liver The liver is firm, enlarged and has a mottled appearance The portal tracts show numerous bile ducts but portal veins are small and scanty Portal pressure 24 mm Hg

(*c*) Patient W M, age 47 Cirrhosis with coarse regeneration nodules and much fibrous tissue Portal pressure 28 mm Hg

(*d*) Patient L R, age 46 Liver very adherent with thick fibrous capsule and normal parenchyma underneath it Portal pressure 22 mm Hg


(a)


(b)


(c)


(d)

Fig 12




(a)




(b)




(c)




(d)

Fig 13

Fig 13 Gross and microscopic appearances of the liver in cases of cirrhosis showing the variations in type Photographs and biopsies taken at operation Histology—Mallory stain × 30

(a) The fibrous tissue is confined to the portal tracts and their immediate neighbourhood Portal pressure 36 mm Hg

(b) The fibrous tissue is distributed as fine strands or septa linking the portal tracts Portal pressure 25 mm Hg

(c) The fibrous tissue is more extensive but the lobular pattern is still recognisable and the surface of the liver is not grossly irregular Portal pressure 21 mm Hg

(d) The fibrous tissue is extensive and the liver parenchyma occupies isolated islands in which the normal lobular pattern is unrecognisable there has been much regeneration and the surface presents a more nodular appearance Portal pressure 27 mm Hg

It is now four years since another of these children, described on p 96, had a portacaval anastomosis, and she has remained in excellent health since

In about half the cases of cirrhosis of the liver which are referred because of evidence of portal hypertension no definite etiological factor can be assigned It is important, however, to appreciate that in many of these the pathological process in the liver is static and that often cirrhosis is not necessarily a progressive disease leading inevitably to fatal liver failure This applies particularly to those cases which are the result of virus hepatitis The inflammatory process has led to proliferation of connective tissue cells, these become converted into fibrous tissue some regeneration of liver cells takes place, and a liver which can function normally remains The only abnormality which endangers the patient is the increased resistance to the flow of blood through the organ

The pattern of fibrosis

The fibrosis in the liver shows wide variations and owing to the regeneration it is not always easy to recognise the particular pattern Occasionally the fibrous tissue is confined to the portal tracts and their immediate neighbourhood, and the liver parenchyma is normal throughout most of the individual lobules, the architecture of the liver being little distorted (Fig 13a) In such cases the obstruction is presinusoidal and functionally the condition will behave like a pre- or extra-hepatic obstruction, in this type the liver function is good and will remain unimpaired unless there is a change in the pathology of the liver More commonly the fibrous tissue is distributed as fine strands or septa linking up one portal tract with its neighbours and thus tending to isolate the liver lobules (Fig 13b), when this pattern obtains the liver retains its normal shape and size, the surface will be only slightly less smooth than normal and the consistency only a little more firm Casual palpation of the liver at a laparotomy may lead to the pathology being overlooked, though this is a type in which the portal pressure is often very high Some of the cases of this type are undoubtedly a sequel to an attack of virus hepatitis In a few cases this type is progressive and gross fibrosis and liver failure supervene, but in the majority the condition is stationary and liver function may remain normal for many years

Grosser forms of cirrhosis are commonly the result of nutritional deficiency or of alcoholism, which have led to necrosis and post necrotic scarring (Fig 13c and d) In this type, with much fibrous tissue the portal pressure is not usually so high, but when large regeneration nodules are seen (Fig 12c) the pressure tends to be higher, possibly because the regeneration nodules distort and compress the portal tracts These cases are often fatal as a result of liver failure, and post-mortem studies by means

of injection casts have shown that there is gross distortion of the portal tracts (Maddon *et al* 1954)

There have been a few cases of portal hypertension with no evidence of extra hepatic obstruction in which the liver pathology has shown only the slightest variation from the normal pattern and the suggestion is made that vascular spasm may play a part in leading to the obstruction of



Fig 13 Liver histology from a patient with no extra hepatic obstruction and a portal vein pressure of 25 mm Hg There is no fibrosis in the liver × 100

blood flow through the liver In this group the liver is a little firmer in consistency than normal rather darkly coloured but the surface smooth or only slightly irregular and microscopically there is no increase in fibrous tissue the portal tracts appear normal or show only a little cellular infiltration Three patients who fall in this group have done well after portacaval anastomosis The histology of a portal tract in one of these cases is shown in Fig 14 The following is one example

J C male age 29 had four severe haematemeses during six years His portal pressure was 27 mm Hg and fell to 6 mm Hg after a portacaval anastomosis he has remained well for six years since that operation His liver biopsy showed no excess fibrosis or cellular reaction in the portal areas

The capsule of the liver

Another pathological type of liver which is associated with portal hypertension is one in which the capsule is greatly thickened and may even be calcified in parts there are two examples in this series one a case of haemochromatosis showing much calcification is described on p 44 The other is noted here

L R male had at the age of 39 an illness with ascites for which paracentesis was performed once and the fluid never recurred Seven years later he had a haematemesis for which he was transfused with four pints of blood His spleen was not palpably enlarged but he had extensive oesophageal varices Liver function tests were almost normal At operation the liver was densely adherent to surrounding structures but when these were freed the capsule was thickened and white (Fig 12d) On incision this capsule was 4 mm thick but beneath this the liver substance looked normal The portal vein was exposed and was healthy except for a small mural thrombus which was removed the pressure in it was 22 mm Hg An end to side portacaval anastomosis was performed

The explanation of this thickened capsule is difficult but it may represent the end result of extensive peripheral necrosis in the liver somewhat analogous to cortical necrosis in the kidney, Daniel s findings in animals (p 5) suggest the idea that sometimes the periphery of the organ is deprived of blood The finding of traces of portal tracts in this fibrous capsule suggests that it does in fact represent a replacement of liver parenchyma by fibrous tissue

Hepatic vein obstruction

A particularly interesting form of intra hepatic obstruction to the portal blood flow is seen when the primary lesion is in the hepatic veins and thus the obstruction is post sinusoidal Two distinct types are seen veno-occlusive disease of the liver which affects the smaller tributaries of the hepatic veins and Budd Chiari disease in which the larger hepatic veins and particularly their orifices into the inferior vena cava are affected Clinically the two conditions have some resemblance Thus hepatomegaly and ascites are prominent features at first the spleen is not greatly enlarged and liver function is not much impaired If recovery from the acute phase takes place fibrosis which commences at the centre of the lobules occurs in the liver and signs of portal hypertension become evident increasing enlargement of the spleen and haematemesis from oesophageal varices Death may be due either to liver failure or haemorrhage from the varices Stuart and Bras (1957) have given a full account of veno occlusive disease as it occurs in Jamaica and suggest that the

ingestion of certain herbs, especially Senecio (ragwort) and Crotalaria (rattle-box) may be important aetiological factors The following is a typical case of Budd-Chiari disease

J H, male, age 21, noticed swelling of his abdomen which gradually increased, after eight months, pain which was never severe was present in the right hypochondrium and he had a little vomiting and epistaxis On admission to hospital a year after the first symptoms there was no jaundice, his abdomen was greatly distended by ascites, there were numerous spider naevi, but no sacral or ankle oedema After paracentesis a hard liver edge was palpable and the spleen could just be felt Extensive varices were present in the oesophagus Liver function tests on these occasions were all within normal limits

At a laparotomy the liver was hard and nodular, the spleen about three times normal size and the portal venous pressure was 19 mm Hg A large vein was present in the falciform ligament Liver biopsy showed severe centrilobular necrosis and haemorrhage with enlargement of the sinusoids (Fig 15) Following a haematemesis he died in hepatic coma

At the autopsy much blood was present in the alimentary canal, the liver weighed 2070 gm and was nodular, the portal vein was healthy All the orifices of the hepatic veins into the vena cava except one were completely occluded by organised membranes, and the remaining one had an opening only 3 mm in diameter (Fig 16), in the liver the hepatic veins were dilated In the vena cava was a little organised thrombosis on the wall, and it must be assumed that the membranes occluding the orifices of the hepatic veins were the result of old thrombi It was surprising that his liver function was so little impaired when there was practically no outlet from the liver by the hepatic veins

It has been shown by Popper and his colleagues (1952) by injection studies of the liver in patients who have died of cirrhosis that the fibrous tissue contains a network of vessels which communicate with both the portal and hepatic veins No such communications were found in normal livers It thus appears that where there is extensive fibrosis in the liver intra-hepatic portal-systemic venous anastomoses develop, and it is suggested that these may go some way towards reducing the portal blood pressure This might account for the fact that when the liver is grossly scarred by fibrous tissue the risk of haemorrhage is less than in those patients with only thin incomplete fibrous septa

The appearance of the liver when exposed at operation may thus show a wide variation, it may look almost normal, the surface may be finely granular when there is an even distribution of fine strands of fibrous tissue throughout When the fibrous tissue is less evenly distributed and there are small nodules of regeneration the typical "hob-nail" appearance is found In a few cases large nodules may push their way above the surface, the intervening capsule being tethered down by fibrous tissue, and here an extraordinary nodular appearance is presented (Fig 17) In the majority of these the etiology is obscure but very coarse liver nodules have been recorded in association with fibro cystic disease of the pancreas

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(a)



(b)

Fig 15 Budd Chiari syndrome Liver biopsy shows centrilobular necrosis and haemorrhage (a) × 30, (b) × 100

There are other findings at operation, the significance of which is not entirely clear, when ascites is present, and sometimes in other cases, oedema may be found between the folds of the lesser omentum and perhaps extending to the adjacent duodenum and retroperitoneal tissues In other cases, soft vascular enlarged lymph nodes are present around the portal vein while in others enlarged lymphatic vessels may be seen in this situation or on the surface of the liver or diaphragm (Fig 18) These vessels normally contain clear lymph but occasionally chyle flows when



Fig 16 Budd Chiari syndrome The inferior vena cava has been opened from behind all the orifices of the hepatic veins are occluded except one which is reduced to 3 mm diameter



Fig 17 Liver with very coarse regeneration nodules in a boy aged 12 Portal pressure 31 mm Hg This patient died of liver failure five years after a porta caval anastomosis



Fig 18 A large lymphatic vessel on the pleural surface of the diaphragm in a case of cirrhosis without ascites

one is divided in the free margin of the lesser omentum, suggesting that in such a case the flow of lymph was upwards, though not necessarily into the liver

Mechanism of ascites formation

Clinical experience shows that portal hypertension *per se* does not cause ascites Cases of primary extra hepatic obstruction, many of whom have portal pressures greater than 30 mm Hg, have no ascites Ascites is, however, common in patients with intra-hepatic obstruction Evidence is gradually accumulating (Madden *et al*, 1954) to show that ascites is due to obstruction of the liver outflow This may occur in cardiac cirrhosis, in which centrilobular fibrosis is found, it also occurs in the Budd-Chiari syndrome and in veno-occlusive disease of the liver, in both there being obstruction to the hepatic veins In these conditions the obstruction to the blood flow is post sinusoidal, and ascites is a prominent feature As a contrast, in those cases in which the fibrosis is confined to the periportal areas and the pressure is high, ascites is as a rule absent There are intermediate cases, in which the architecture of the liver is quite altered by regeneration, and in which the obstruction is both pre- and post sinusoidal, and then ascites is associated with a raised portal pressure However, when there is very extensive fibrosis intra hepatic portal systemic shunts may help to bring down the portal pressure, and this would account for the fact that cases in which ascites is a prominent feature do not often have severe haemorrhages from oesophageal varices

There are undoubtedly other factors in the formation of ascites and changes in the plasma proteins which occur in liver disease certainly play a part In patients who have had haemorrhages from oesophageal varices and who also have ascites the latter symptom may not reappear after a successful portacaval anastomosis which suggests that when there is defective liver function the ascites which is already present may be aggravated by the raised portal pressure

REFERENCES

CHILD C G ... and O NEILL ... Portal Vein ... *...ast* 94 31 45

DIBL ... ation Necrosis and Fibrosis in the Liver *... n med J* 1 833 841

GELFAND M (1957) *Personal Communication*

GIBSON J B and RICHARDS R. I (1955) Cavernous Transformation of the Portal Vein *J Path Bact* 70 81–96

HIMSWORTH H P and GLYNN L E (1944) Toxipathic and Trophopathic Hepatitis *Lancet* 1 457 461

HUNT A H and WHITTARD B R. (1954) Thrombosis of the Portal Vein in Cirrhosis Hepatis *Lancet* 1 281 284

LEATHER H M (1957) *Personal Communication*

PARKER R A and SEAL R M E (1955) Cavernous Transformation of the Portal Vein *J Path Bact* 70 97 103

PARKER R G (1956) Fibrosis of the Liver as a Congenital Abnormality *J Path Bact* 71 359 368

POPPER H ELIAS H and PETTY D E (1952) Vascular Pattern of the Cirrhotic Liver *Amer J clin Path* 22 717–729

REICHMAN S and DAVIS W D (1957) The Splenic Approach to the Portal Circulation *Gastroenterology* 33 609–615

SHALDON C (1958) Portal Pyaemia *Brit J Surg* 45 357 360

STUART K. I and BRAS G (1957) Veno occlusive Disease of the Liver *Quart J Med* 26 291 315

CHAPTER III

EFFECTS OF PORTAL HYPERTENSION

Portal hypertension produces anatomical and functional changes in the splanchnic system these are mainly associated with the development of the collateral circulation and the congestive splenomegaly

In long standing cases the visceral peritoneum loses its normal lustre and presents an opaque grey appearance The dilated veins of the portal system show changes similar to varicose veins in the legs in places the wall is thinned and mural thrombi are likely to be present such veins tear easily *and do not hold sutures well In cases of intra hepatic obstruction* other findings may be present the lymph nodes in the hilum of the liver and along the portal vein may be enlarged and sometimes there is oedema of the neighbouring structures particularly the gall bladder the lesser omentum and the adjacent retroperitoneal tissues These are probably the result of the liver disease rather than being directly due to the portal hypertension

The collateral circulation

This occurs where the portal and systemic circulations join (Figs 19 and 20)

The oesophagus and gastric fundus The most important site is the region of the oesophagus and the fundus of the stomach (Fig 21a b and c) here the varices may be very extensive often extending in the sub mucosa of the oesophagus up to the level of the arch of the aorta and even as high as the junction of the pharynx with the oesophagus (Fig 22) their actual arrangement shows considerable variation there may be one or two large trunks or a network of irregular channels which lie all round the circumference When viewed through an oesophagoscope intense purple congestion of the whole mucosa of the lower part may be seen but in some cases a few large vessels stand out projecting into the lumen either as isolated swellings or running for some length upwards producing exag gerated longitudinal folds as in Fig 23 usually the blue colour of the con tained blood can be seen through the wall but sometimes the overlying epithelium is thickened and the varices appear grey When a thoracotomy is carried out dilated veins are usually seen on the outer surface of the oesophagus and the points where they penetrate the muscular coat may be clearly visible At the same time it may be noted that there are dilated veins on the upper surface of the diaphragm, in addition the pericardio

phrenic veins and one or two of the lower intercostal veins may be enlarged (Fig 59)

The retroperitoneal tissues In the retroperitoneal tissues the most constant collateral veins are found in the neighbourhood of the spleen thus the vasa brevia are often tremendous carrying blood up from the spleen towards the oesophageal varices in the lieno renal fold of peritoneum the collaterals usually take the form of many small channels each not more than one millimetre in diameter but sometimes a single large

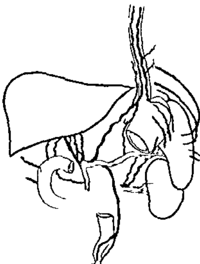


Fig 19 The collateral circulation in extra hepatic obstruction due to thrombosis of the portal vein

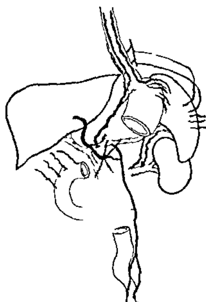


Fig 20 The collateral circulation in intra hepatic obstruction

vessel is found running from the lower pole of the spleen into the retroperitoneal tissues and such a vessel will sometimes show on a venogram (Fig 22) When performing a splenorenal anastomosis the left adrenal vein is often found to be dilated and varicose

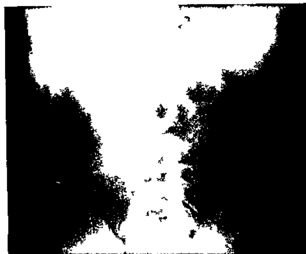
These retroperitoneal veins drain into the renal veins (Fig 25) the lower *intercostal veins and by the latter to the azygos and hemiazygos veins* On more than one occasion when approaching the oesophagus by a left thoracotomy the hemiazygos vein has been found to be more than one cm in diameter and either this vein or the azygos vein may show up as paravertebral shadows on X ray films of the chest (Fig 26)



(a)



(b)



(*c*)

Fig 21 Varices in the gastric fundus showing (*a*) as filling defects when the stomach contains barium and (*b*) the veins outlined by trans splenic venography A case of extra hepatic obstruction with thrombosis of the splenic vein (*c*) The venogram by injection into a jejunal vein shows that the portal vein is patent



(a)



(b) (c)

Fig 22 Oesophageal varices extending as high as the cervical oesophagus in a case of intra hepatic obstruction (same patient as Fig 13a)

Fig 23 Oesophageal varices presenting as a few large vessels running up the oesophagus

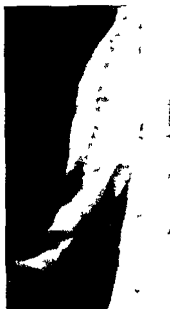
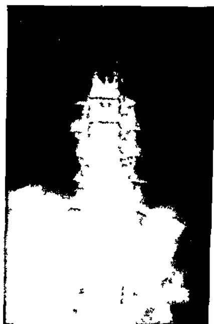




Fig 24 Venogram in a case of extra hepatic obstruction a large collateral vessel encircles the lower pole of the spleen and drains to the chest wall by the lieno renal ligament



Fig 25 Trans splenic venogram in a case of extra-hepatic obstruction, showing retroperitoneal collateral vessels leading to the left renal vein and the inferior vena cava The filling of the latter shows through the gastric translucency, the stomach being displaced by the large spleen





(a) (b)

Fig 26 (a) Large hemiazygos vein showing as a paravertebral shadow The patient had had an end-to-side spleno-renal anastomosis ten years before using a vitallium tube, which can be seen overlying the twelfth rib

(b) Tomogram of the same patient showing the shadow of the enlarged azygos vein above the left branchus

Following an operation collaterals develop through adhesions to the scar (Fig 27)

The anal canal In the anal canal internal haemorrhoids are often found associated with portal hypertension but to what extent this is cause and effect it is difficult to say, the majority of cases of cirrhosis of the liver *occur in patients who are getting* past middle age and this is the age when haemorrhoids are common in children who have portal hypertension haemorrhoids do not occur but some congestion of the rectal and anal mucosa may be present *It is doubtful whether serious* bleeding from haemorrhoids is any more common in patients with portal hypertension than in other patients who have haemorrhoids and are in the same age group

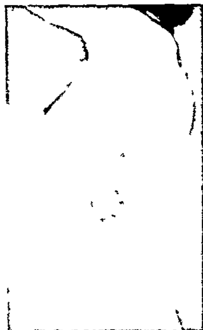


Fig 27 The same patient as shown in Fig 26 *infra red photograph* Dilated subcutaneous veins around the scar of the operation

The falciform ligament Mention has already been made of the collateral circulation in the falciform ligament, here again it may take the form of a single large channel (Fig 28) but more often it consists of a mass of tiny vessels *whose appearance can be readily recognised during a peritoneoscopy* From the abdominal attachment of the falciform ligament these vessels pierce the linea alba and thus give rise to the dilated veins in the subcutaneous tissues of the abdominal wall forming the 'Caput Medusae These vessels may be troublesome when making an upper abdominal incision When there are large veins in the falciform ligament and one of them pierces the linea alba through a narrow opening a sub*cutaneous varix may form at this point and the flow through this will set* up currents which cause a palpable thrill and a murmur which can be heard with the stethoscope the condition being described as the Cruveilhier Baumgarten syndrome it is not necessary to postulate an abnormal anatomical arrangement for such a vein simply represents a persistent vein of Burow or possibly a persistent umbilical vein patent throughout its course



Fig 28 Trans splenic venogram in a case of intra hepatic obstruction showing a large vein coming from the left branch of the portal vein and traversing the falciform ligament

Cruveilhier Baumgarten syndrome

W J , male, first noticed some varicose veins near his left groin at the age of 14 and these gradually spread upwards When aged 26 he had some melaena, haematemeses started the following year and were repeated almost every year until he reached the age of 37

His liver was just palpable, the spleen was enlarged to three fingers and varices were present in his oesophagus The dilated tortuous vein in the abdominal wall extended from the pubes to a point just above the xiphisternum Five cm above the umbilicus was a varix 4 cm in diameter in which

bromsulphalein test showed 16 per cent retention after 45 minutes

An end to side portacaval anastomosis was performed The large vein in the falciform ligament was a branch of the left portal vein There was some mural thrombosis in the main portal vein and the more recent part of this clot was removed The pressure in the portal vein was 15 mm Hg while that in the vein in the falciform ligament was 14 mm Hg and fell to 3 mm Hg

on compression of the portal vein Before the anastomosis the pressure in the inferior vena cava was 7 mm Hg and that in the portal vein fell to this level after the anastomosis was open

Since the operation he states that he has felt better than he had done for years there has been no more haematemesis and he has for five years worked consistently as a welder in the docks The vein in the anterior abdominal wall thrombosed a few days after his operation and soon after became invisible (Fig 30)

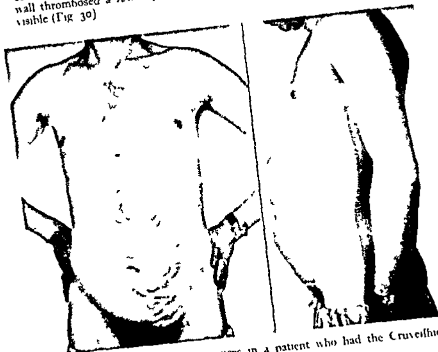


Fig 29 Large subcutaneous varices in a patient who had the Cruveilhier Baumgarten syndrome

Some of the largest collateral channels are seen in the omentum which has become adherent to some structure drained by the systemic system [illegible] ch a vein in the omentum which was adherent to the lateral [illegible] m from a patient in whom a large [illegible] s and [illegible] ,7) to one ovary

Occasionally varices develop in other sites, thus [illegible] record the case of a patient with cirrhosis of the liver who had severe bleeding from a varix in the caecum which necessitated an emergency right hemicolectomy but the patient died of liver failure

Fatal intraperitoneal haemorrhage has also been recorded by Ellis Griffiths and MacIntyre (1958)

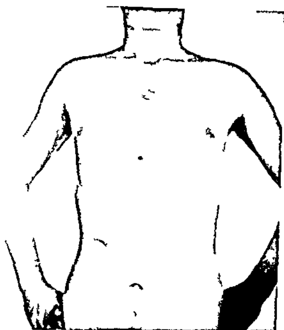


Fig 30 The same patient as in Fig 29 a few months after a portacaval anastomosis



Fig 31 Photograph taken at operation showing a vein 2 cm diameter in the omentum which was adherent to the lateral abdominal wall the patient had both intra and extra hepatic obstruction

It is difficult to give an explanation of the fact that in some patients one group of collaterals predominates and in others another group It has [illegible]

become insignificant, it seems that very much the same takes place in cases of portal hypertension for one patient may have very large oesophageal varices and little dilated collateral circulation elsewhere, while another will have a large group of collaterals in another site and few if any



Fig 32 Trans splenic venogram showing a large collateral vein in the omentum which was adherent to the wall of the left side of the pelvis In spite of this anastomosis the pressure in the portal vein was 20 mm Hg and the patient had many serious haemorrhages The portal vein is narrowed by mural thrombus

dilated veins in the oesophagus The same applies to the splenic vein in some cases particularly those with marked splenic enlargement this vein will be greatly dilated and somewhat tortuous but in other cases this vein may not be dilated at all

The spleen

The spleen is always enlarged in cases of portal hypertension, and it is unusual for it not to be palpable However, there is little or no relationship between its size and the portal blood pressure That its enlargement is partly the result of congestion is shown by the fact that it will become considerably reduced in size within a few hours of a portacaval anastomosis But long standing congestion causes structural changes in

the spleen and these take a long time to resolve after the hypertension has been relieved In addition, the work of Cameron and de Saram (1939) suggests that in disease of the liver there may also be a blood borne factor which causes enlargement of the spleen, for they found in rats that when the spleen was transplanted into the abdominal wall and was entirely cut off from the portal circulation, it would still show some enlargement when cirrhosis of the liver was induced by means of carbon tetrachloride

Hyperspleniam The enlarged spleen can give rise to changes in the blood, thus there may be anaemia, leucopenia or thrombocytopenia It is sometimes difficult to determine whether the anaemia is due to haemorrhage or to an effect of the spleen on the bone marrow, and probably there is often a combination of both factors The thrombocytopenia may lead to nose-bleeding, bruising or petechiae of which the patient may complain Regarding the white cells, a leucopenia of about two to three thousand is common but this does not seem to have serious clinical significance After a portacaval anastomosis, as the spleen becomes reduced in size, the blood picture tends to return to normal, but sometimes the blood changes continue to be troublesome, only once in our series (a case of haemochromatosis) was it necessary, after the establishment of a portacaval anastomosis, to perform a splenectomy because of a persistent haemolytic anaemia

M F , female, at the age of 45, had three severe haematemeses within the space of a month necessitating transfusion of 12 pints of blood or plasma. She had [illegible]

Her leucocytes fell at one stage to 600 per cu mm and platelets were scanty After a portacaval anastomosis, at which it was confirmed that the calcification was entirely in the thickened capsule, the blood picture improved to normal However, a year later she developed a severe haemolytic anaemia, which was only checked by splenectomy At this operation the typical red brown colour of the pancreas found in cases of haemochromatosis was noted Following this she was very fit for three months, then she became mentally confused, losing her muscular power, and died after a further three months

REFERENCES

CAMERON, G R and de SARAM, G S W (1939) "A Method for Permanently Dissociating the Spleen from the Portal Circulation (The 'Marsupialised' Spleen) and its Use in the Study of Experimental Liver Cirrhosis" *J Path Bact*, 48, 41–47

ELLIS, H, GRIFFITHS, P W W and MACINTYRE, A (1958) "Haemoperitoneum." *Brit J Surg*, 45, 606–610

LEVY, J S, HARDIN, J H, SHIPP, H and KEELING, J H (1957) "Varices of Caecum as an Unusual Cause of Gastrointestinal Bleeding" *Gastroenterology*, 33, 637–640

LONGLAND, C. J (1953) "The Collateral Circulation of the Limb" *Ann R Coll Surg*, 13, 161–176



(*a*)



(*b*)

Fig 33 Calcification in the capsule of the liver in a case of haemochromatosis

(*a*) The trans splenic venogram which in addition to the calcification shows large varices around the cardia and an unusually vertical portal vein

(*b*) Lateral view showing the calcification which was shown at operation to be confined to the capsule

CHAPTER IV

CLINICAL ASPECTS OF PORTAL HYPERTENSION

The only clinical findings due directly to portal hypertension are those associated with the collateral circulation, or the effects of the enlarged spleen Patients with intra hepatic obstruction may have in addition clinical features due to their liver disease

The oesophageal or gastric varices

When these extend a long way up the oesophagus the patient may feel a sense of fullness in the throat, but the serious aspect is the danger of haemorrhage from these varices Occasionally it manifests itself as a slow ooze, so that the patient is unaware of it, but suffers from anaemia and occult blood is found in the stools More often the bleeding is sudden and severe, in Great Britain such bleeding accounts for less than ten per cent of cases of serious haematemesis, the great majority being due to peptic ulceration but there are parts of the world such as Central Africa where this state of affairs is reversed, and in the great majority of cases of haematemesis the bleeding comes from oesophageal varices However, in children, even in Britain, portal hypertension is the most common cause of haematemesis The actual precipitating factor may be peptic ulceration of the oesophageal mucosa over the varix, but trauma by food particles has also been blamed, and there may be other factors such as a sudden rise in the intra-abdominal or portal pressure Krook (1956) has shown that the Valsalva manoeuvre can increase the occluded hepatic vein pressure to as much as 100 mm Hg in patients with or without cirrhosis and considers sudden abdominal strain is a likely cause of rupture of varices However, the intervals between one haemorrhage and the next can be very variable, anything from a day or two to many years It is this unpredictability which makes the outlook in these cases so uncertain Some figures show (Patek *et al*, 1948) that of patients with cirrhosis of the liver, more than half are dead within a year of their first haemorrhage, but some of these die of liver failure and not all of them succumb to another attack of bleeding Contrary to the experience of some writers, we have found it very unusual for a patient to die as a result of his first haemorrhage, but the immediate prognosis for a patient bleeding from oesophageal varices is more serious than in cases of bleeding from peptic ulceration Usually the blood is vomited without any warning at all, though the patient sometimes notices a salt taste in the mouth before the blood appears In those

cases in which the blood all passes downwards, the patient may feel faint, and the appearance of black stools follows A second severe haemorrhage within a few days is particularly serious

Following a haemorrhage the patient may pass into a state of liver failure, or may suffer from neuropathic symptoms and even pass into coma as a result of the absorption of protein products resulting from the digestion of the blood in the intestine (see Chapter X)

The spleen

The spleen is always enlarged to some extent in cases of portal hypertension It is not always palpable, for it may not extend below the costal margin, or the presence of ascites may prevent its being felt In many cases it is enlarged as much as four fingerbreadths below the costal margin, but it is unusual to find it much larger than this As already mentioned, the size of the spleen bears little or no relationship to the elevation of portal pressure, but it is proportionately more enlarged in young patients than in those in the older age group As a rule it is free of adhesions, but sometimes, probably as a result of infarction adhesions to the parietal peritoneum are present and in that case they will be very vascular

As a result of haemorrhage the patient may be anaemic, but this may be in part due to haemolysis, the effect of hypersplenism, there may be petechiae or echymoses, and nose bleeding is a not infrequent symptom Some patients merely complain of lassitude

Veins on the abdominal wall

The presence of dilated veins in the subcutaneous tissues of the anterior abdominal wall which tend to show a radiating pattern from the umbilicus has already been mentioned as a sign of intra hepatic obstruction (Fig 29) [illegible]

hypertension, there may be evidence of associated disease, particularly of the liver Thus ascites, palmar erythema spider naevi (Fig 34), gynaecomastia and testicular atrophy are all evidence of liver disease The patient may have the characteristic brown complexion of the cirrhotic, or there may be definite jaundice This last is a sign that there is either some biliary obstruction or that the hepatitis is in an active phase, and adds greatly to the danger of any treatment for the hypertension, in fact it is wise to postpone any active treatment of the latter until at least a few months after the jaundice has subsided, unless delay is dangerous for other reasons

The liver itself is sometimes palpable and this is an indication that intra-hepatic obstruction is present When the liver can be felt in the epigastrium it does not necessarily mean that it is enlarged, for a liver of

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Veins on the abdominal wall

The presence of dilated veins in the subcutaneous tissues of the anterior abdominal wall which tend to show a radiating pattern from the umbilicus has already been mentioned as a sign of intra hepatic obstruction (Fig 29) Similar veins may be noted in the region of a scar of a previous operation to the deep aspect of which there are adhesions (Fig 27)

In addition to these signs which are the direct result of the portal hypertension there may be evidence of associated disease particularly of the liver Thus ascites palmar erythema spider naevi (Fig 34) gynaecomastia and testicular atrophy are all evidence of liver disease The patient may have the characteristic brown complexion of the cirrhotic or there may be definite jaundice This last is a sign that there is either some biliary obstruction or that the hepatitis is in an active phase and adds greatly to the danger of any treatment for the hypertension in fact it is wise to postpone any active treatment of the latter until at least a few months after the jaundice has subsided unless delay is dangerous for other reasons

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normal size which is unduly hard will be palpable In the hepatic con ditions which cause portal hypertension the liver may in fact be *smaller* than normal or considerably enlarged but in the *majority of cases it is of* about average size When it is much enlarged this again is usually an indication that there is still some active hepatitis

Ascites unless attributable to some quite independent cause *is evidence* that the obstruction is intra hepatic though there may of course be secondary portal vein thrombosis I have only seen one patient with



F g 34 Unusually extensive sp der naevi on the shoulder of a pat ent th alcohol c c rrhos s of the l ver

ascites extra hepatic obstruction and no disease of the liver but in th s case there were probably other factors which gave rise to the intra peritoneal fluid and it is my bel ef that extra hepatic obstruct on by itself never causes ascites

REFERENCES

KROOK H (1956) C rculatory Stud es in Liver Cirrhos s *Acta Med Scand Suppl* 318 60

PATEK A J POST J RATNOFF O D MANKIN H and HILLMAN R W (1948) D etary Treatment of Cirrhos s of the Li er *J Amer med Ass* 138 543 548

CHAPTER V

MANAGEMENT OF PORTAL HYPERTENSION

In the management of a patient with portal hypertension, the first steps are the establishment of the diagnosis, the decision that the hypertension is the cause of the patient's symptoms, and whether it is sufficiently severe to endanger his life or seriously to impair his health. In cases of intra-hepatic obstruction it is important to obtain a clear distinction between those symptoms due directly to the disease of the liver, and those which are a consequence of the hypertension.

Diagnosis

The great majority of patients who have portal hypertension which require treatment present as a result of haemorrhage from the alimentary canal usually haematemesis. Sometimes they go to the doctor because of malaise, and occasionally the patient himself will have noticed the swelling due to his enlarged spleen. The presence of these findings will lead the doctor to suspect portal hypertension, but in cases of sudden haemorrhage, the distinction from peptic ulcer may be difficult, and a number of cases are only diagnosed when an emergency laparotomy for haematemesis is performed. However, in the great majority the finding of the enlarged spleen will be the most significant sign. There may be a history of past jaundice and other signs of liver disease.

After the clinical examination a barium swallow should be ordered to demonstrate the presence of varices, but the radiologist should also look for any other abnormality such as a peptic ulcer in the stomach. When varices are present in the oesophagus or stomach they show as rounded filling defects projecting into the lumen (Fig. 22), but if too much barium is swallowed at a time it may compress and obliterate the varices and their presence may be missed. There are, however, a few patients in whom the varices fail to show as a result of this radiological examination, and if the diagnosis is seriously in doubt these patients should be examined with an oesophagoscope, by which means it is unlikely that any varices will be overlooked. The varices show up as rounded prominences in the oesophageal mucosa, sometimes up to 1 cm. in diameter, and tending to run longitudinally; normally the blue colour of their contained blood shows through the thin mucosa, but sometimes they have an opaque grey colour where thickening of the epithelium has occurred over them. Tanner (1958) considers that they can be seen better with a gastroscope as it is

withdrawn up the oesophagus than with the oesophagoscope, gentle inflation being applied at the time

For practical purposes the presence of varices may be taken as definite evidence of an appreciable degree of portal hypertension, and the reverse also holds good As mentioned earlier, in some patients the collateral circulation may be much more developed in one site than another, but as the most serious consequences of the hypertension result from the varices about the cardia, the absence of varices in the oesophagus suggests that there is not a sufficient degree of hypertension to require treatment

While these investigations are being undertaken, certain pathological tests should be made A full blood count is required, which should include differential leucocyte and platelet counts This will indicate any anaemia as well as the effects of hypersplenism In difficult cases a bone-marrow examination may be necessary Liver function tests are also required Laboratories differ in the tests that they carry out as a routine, but it is essential to have estimations of the serum bilirubin and alkaline phosphatase, and a differential protein estimation, either chemically or by electrophoresis, with the absolute value of the serum albumin is required At least one of the flocculation tests should be done, preferably the thymol turbidity, as these give some indication of activity of liver disease In some clinics much weight is attached to the bromsulphalein excretion test, and this is of value in patients whose liver function is impaired and the question of a major operation is being considered

Peritoneoscopy (Walker, 1943) and liver puncture biopsy are sometimes helpful in making the diagnosis, the former has its greatest use if there is a possibility of malignant disease in the liver, either primary or secondary, and can be carried out under local anaesthesia Liver biopsy by needle puncture is occasionally indicated if there is doubt concerning the nature of any pathological process in that organ It should only be done if the prothrombin time is normal As there may be large vessels in the falciform ligament the approach through the epigastrium should be avoided in these cases The method is not without risk (Terry, 1952) and should only be employed if the decision for or against surgical treatment depends on knowledge of the pathology of the liver It is possible to obtain a liver biopsy under vision with the peritoneoscope but by either method the specimen may be too small to give a conclusive diagnosis

Venography

Before any surgical treatment is undertaken it is most important to have a knowledge of the state of the veins of the portal system This will indicate the site of any obstruction, and in fact venography may in some cases be the only way of distinguishing between intra- and extra-hepatic obstruction (Walker, 1957) (Steiner *et al*, 1957)

Except in patients who have had the spleen removed, this should be done in the first instance by percutaneous splenic puncture (Walker *et al*, 1953) For adults only local anaesthesia is required but in children a general anaesthetic should be given The puncture is made either below the left costal margin when the spleen is much enlarged, or else through the intercostal space, usually the ninth, which is in line with the apex of the spleen If the spleen is not palpable the injection should be made over the area of splenic dullness, but it is our practice only to carry out this examination if the spleen is impalpable in those patients in whom the information cannot be obtained in any other way A 17-S W G needle is used, and as a rule the change in consistency can be felt as the needle enters the spleen The patient is instructed to take only very shallow breaths while the needle is in place Before the injection is made it is usually possible to withdraw blood into the syringe, but we have not found this in every case For an adult, 30 ml of a 70 per cent solution of one of the soluble opaque iodine compounds is used, and a proportionately smaller dose for children The injection is made as rapidly as possible, films are exposed as the injection is completed and after an interval of three seconds, though with special casette changers more films can be exposed, but two will give all the information required The needle is withdrawn as soon as the injection is completed While the needle is in place before the injection is given, the opportunity may be taken to record the intra-splenic pressure, using a saline manometer we have had rather inconstant readings, and on one occasion the high pressure indicated that the needle was in an artery It is possible that with a continuous recorder more accurate readings of the intra-splenic pressure can be obtained

Further venographic studies can be made only at a laparotomy, and in fact if the patient has had the spleen removed this is the only way In cases of intra-hepatic obstruction if a percutaneous venogram shows that the splenic and portal veins are unobstructed the case is clearly one of intra-hepatic obstruction, and an anastomosis to either the portal or the splenic vein will provide a suitable by pass When, however, none of the injection reaches the portal vein further study may be necessary, for the injected fluid may be diverted because the vein is obstructed by thrombus, or because the flow of blood is reversed To demonstrate the patency of veins at laparotomy the best site for the injection of the opaque medium is into one of the veins of the mesentery of the small intestine, and at the same time the portal pressure can be measured No attempt at a venous anastomosis should ever be made before it has been shown by venography that the vein at the proposed site of anastomosis is patent, and that it is in such a position that it will effectively drain the portal circulation, particularly the region of the cardiac end of the stomach

In trans splenic venography in a patient with a normal portal circulation the splenic vein appears as a curved vessel about 1 cm in diameter



Fig 35 Trans splenic venogram in a patient with a normal portal circulation



Fig 36 Trans splenic venogram in a patient with cirrhosis of the liver without demonstrable varices the portal veins in both lobes of the enlarged liver are shown

running across the epigastrium (Fig 35) It is joined by the superior mesenteric vein slightly to the left of the mid-line, the blood entering from this vein causing a filling defect which is carried a little way up the portal vein and gradually fades away The portal vein runs upwards and to the right and divides into its two branches

It has been said that there is normally streamlining in the portal vein blood from the splenic vein usually passing into the left branch of the portal vein while that from the superior mesenteric vein passes mainly to the right branch Venography does not bear this out, in normal cases mixing of the opacified and clear bloods seems to be complete in the portal veins and both lobes fill equally (Fig 36), sometimes thrombosis in one branch may prevent filling of an entire lobe (Fig 9)

By the time the injection is completed the smaller branches of the portal vein in the liver are filled and the appearance is that of vessels dividing repeatedly, each division giving rise to two of small calibre Normally the opaque medium flows rapidly through the liver and later plates show a confused pattern of filling of portal and hepatic veins and sinusoids

In cases of portal hypertension some dilatation of the splenic and portal veins may be seen, while the splenic vein is not only lengthened but rendered more tortuous by the shifting of the hilum of the spleen to the right, which occurs as that organ enlarges The flow of blood is retarded and much of the opaque medium enters collateral channels, and in fact in some cases very little may reach the portal vein and the liver, resulting in faint shadows in these sites in spite of the fact that the veins are fully patent In an extreme case there may be a reversal of blood flow in part of the portal circulation and this possibility must be borne in mind

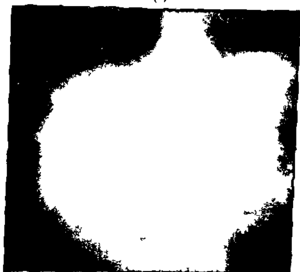
A L , a boy age 7 years had an attack of melaena in June 1956 which was severe enough for him to require a blood transfusion His liver and spleen were found to be enlarged During subsequent months he had repeated haematemeses and blood transfusion His treatment was complicated by a persistent salmonella infection

A year after his first melaena he was transferred with a view to surgical treatment The liver function tests were all normal Intra splenic venography (Fig 37a) showed wide splenic and left gastric veins, but no opaque medium entered the portal vein or the liver When the abdomen was explored a venogram by a jejunal vein (Fig 37b) showed a healthy portal vein, filling of the liver and of a tortuous umbilical vein, though some of the medium injected by this route also entered the left gastric vein and the oesophageal varices His portal pressure was 25 mm Hg and a satisfactory end-to-side portacaval anastomosis was carried out He was a case of congenital fibrosis of the liver

When there is extra hepatic obstruction the site of the commencement of the blocked vein is usually clearly indicated, a mass of collateral vessels surrounds the obstructed segment When the block is in the portal vein only, some filling of the vessels of the liver is usually seen If the splenic



(*a*)



(*b*)

Fig 37 (*a*) Trans splenic venogram in a boy of seven with intra hepatic obstruction No opaque medium reaches the liver as it is all diverted into collaterals especially the left gastric vein

(*b*) Venogram using a jejunal vein in the same patient some of the injection goes by the portal vein *to the liver but the remainder is diverted to the left* gastric vein There is also filling of a vein in the *falciform ligament The portal vein was fully patent* and a portacaval anastomosis was made

and portal veins are obstructed it is unusual for any of the medium to reach the liver but masses of collaterals may be observed around the pancreas and the fundus of the stomach (Fig 21) Sometimes the collateral circulation in the retro-peritoneal tissues is so well developed that some filling of the left renal vein and the inferior vena cava is seen—a natural but inadequate portacaval anastomosis (Fig 25) Occasionally the umbilical vein in the falciform ligament with its origin from one of the two main branches of the portal vein (usually the left) is visible (Fig 28)

Irregularity in the wall of the splenic or portal vein or narrowing of the lumen may indicate partial thrombosis, such a mural thrombosis may not prevent the vein being used for an anastomosis if the lumen is adequate, and it may even be possible to remove some of the clot at the operation In one of our patients the portal vein was found to have a double lumen, old thrombus forming a longitudinal septum in the vein

With the information which has been made available by these investigations the clinician will be in a position to advise on the line of treatment to be carried out on any individual patient Surgical treatment may be required as a life-saving matter when haemorrhage has been serious but there may be strong contra indications to a major operation, or particularly to a venous shunt, and the indications have to be carefully weighed in the balance The succeeding chapters will describe the alternative operative measures which are available and the indications for each, for the main factors in the success of surgery in the treatment of portal hypertension are the selection of the patient and of the appropriate operation

REFERENCES

STEINER, R E, SHERLOCK, S and TURNER, M D (1957) "Percutaneous Splenic Portal Venography" *J Fac Radiol*, 8 158–177

TANNER, N C (1958) *Personal Communication*

TERRY, R (1952) "Risks of Needle Biopsy of the Liver" *Brit med J*, 1 1102–1105

WALKER, R M (1943) "Peritoneoscopy" *Proc R Soc Med*, 36 445–448

—— (1957) "A Review of Trans-splenic Portal Venography in the Investigation of Portal Hypertension, Surgical Aspects" *J Fac Radiol*, 8, 178–180

WALKER R M, MIDDLEMISS, J H and NANSON, E M (1953) "Portal Venography by Intrasplenic Injection" *Brit J Surg*, 40, 392–395

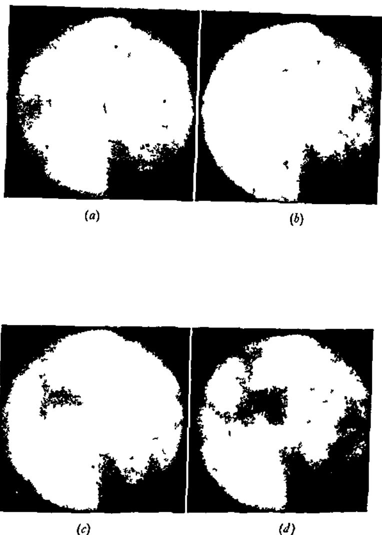


Fig 38 Shots from a cine angiogram taken with an image intensifier Trans splenic venography in a case of intra hepatic obstruction The time interval between each shot was approx 1 second In (a)



(e)



(f)



(g)



(h)

the portal vein is beginning to fill and it is almost clear in (h). From (c) to (h) filling of tortuous collaterals along the lesser curvature of the stomach can be seen. In (g) the portal vein pattern in the liver is becoming blurred by sinusoidal filling.


(a)


(b)


(c)


(d)


(e)


(f)



(g)



(h)



(i)

Fig 39 Shots from a cine-angiogram of another patient, there is a very large and tortuous left gastric vein, which when filled with opaque medium hides the shadows of the vertebrae The portal vein has emptied at (h) but the opaque medium remains in the collaterals much longer

CHAPTER VI

TREATMENT OF PORTAL HYPERTENSION

With very few exceptions, the only indication for surgical measures in the present state of our knowledge is the prevention or treatment of haemorrhage from the alimentary canal However, as ascites is so often associated with portal hypertension, a note on this is not out of place

Treatment of ascites

Some surgeons have advocated operative treatment for the relief of ascites due to cirrhosis of the liver, but it has been pointed out that such ascites is not due to the venous congestion in the portal system though it *may be aggravated by it The presence of ascites due to liver disease* is an indication that there is some defect in the function of the liver Attempts to treat such ascites by surgical means have been made by many surgeons ever since Talma of Utrecht and Morison of Newcastle independently introduced the operation of omentopexy Drainage of ascitic fluid into the urinary tract or back into the blood stream has been given a trial but these all failed as the openings became occluded by adhesions Others have tried to drain the fluid into the tissues of the abdominal wall so that it may pass back into the circulation by the lymphatics, and I have attempted to achieve the same result by denuding much of the anterior abdominal wall of its parietal peritoneum Talc has been applied to the surface of the liver to promote vascular adhesions between the liver and the diaphragm, in view of the evidence that the ascites is associated with obstruction to the outflow from the liver, but the results have not met with striking success Ascites is a variable symptom which may disappear spontaneously, so it is difficult to assess the results of any measures for its relief, fortunately in many *cases it responds well to treatment by a low* salt diet Very few patients can tolerate a diet which is completely salt free, and it is difficult to devise such a diet which is palatable, but it is *possible to reduce the daily intake of sodium to 0 5 gm* and this should be adopted as a first measure when ascites is troublesome Sometimes it has a dramatic effect, in other cases little benefit is obtained Elimination of fluid may be assisted by the use of diuretics such as mersalyl or chlorothiazide

Non-operative treatment of portal hypertension

It has been traditional to treat patients with cirrhosis of the *liver on a* high protein diet and to give glucose by mouth Apart from the calories

that these foods provide there is no evidence that they have any specific effect on the liver Sometimes if the patient has established a good collateral circulation, the high protein may precipitate the onset of neurological symptoms, which may even proceed to coma

Apart from the value of a low salt diet and the use of diuretics in the treatment of ascites and abstention from alcohol, I am therefore drawn to the conclusion that there are no medical measures which influence the course of the disease in Laennec's cirrhosis of the liver, and there is certainly no medicinal treatment which relieves patients with extra-hepatic obstruction Iron should, of course, be given for anaemia which is the result of haemorrhage

When there is a specific cause of fibrosis of the liver it is a different matter Many patients in whom there is an alcoholic factor in the etiology will benefit when they give it up In fact the prognosis of patients with alcoholic cirrhosis seems much worse than those who have cirrhosis following virus hepatitis, and patients with the former condition who have an operation for portal hypertension should be advised of the utmost importance of their giving up their alcohol consumption Schistosomiasis should be treated by the specific drugs advocated for this condition Hepatolenticular degeneration may be slowed in its course by stimulating the elimination of copper

[illegible]

hypersplenism has in the condition Whichever the cause, however, blood transfusion plays an important part and is frequently needed as an emergency for acute haemorrhage, and as a preliminary to operative treatment

Operative treatment of portal hypertension

Surgical treatment can take three forms The first aims at reducing the portal venous pressure to a safe level, which can only be done by

[illegible]

off the flow of portal blood to these vessels The third reduces the blood flow through the liver

1 *Means of lowering the portal pressure* The nearer the by pass is to the site of obstruction the more efficiently will it function, thus if the obstruction is in the liver a direct anastomosis between the portal vein and the inferior vena cava will give the best results It, however, the portal vein is obstructed, the only other vein in the portal system which is large enough is the splenic vein, the nearest available vein in the systemic system being the left renal vein A splenorenal anastomosis has the

disadvantage that the flow in the splenic vein will have to be reversed, and the operation entails removal of the spleen and consequently some of the already formed collaterals in the peritoneal folds of the spleen The tail of the pancreas may distort the splenic vein when the latter is brought down to the renal vein In our experience, splenorenal anastomoses have not had the success which is so striking from the point of view of preventing haemorrhage after a portacaval anastomosis

Other veins of the portal system have been employed and autogenous *vein grafts have been inserted (Rousselot, 1952)*, but such procedures are very likely to fail as a result of thrombosis and it is doubtful whether they are worth attempting

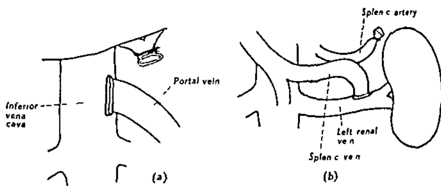


Fig 40 Portal systemic anastomoses which at present have a place in the treatment of portal hypertension (*a*) End to-side portacaval anastomosis (*b*) End-to side splenorenal anastomosis associated with splenectomy

2 *Palliative operations* The palliative measures used which do nothing to bring down the portal pressure, are as follows

(A) Methods which obliterate the varices

- (*a*) Injection through an oesophagoscope of sclerosing solutions
- (*b*) Oesophagotomy and ligature of the veins

(B) Methods intended to interrupt the flow of blood through the varices

- (*a*) Gastric transection
- (*b*) Oesophageal transection
- (*c*) Devascularisation of the region of the cardia

(C) Excision of the varix-bearing area

Oesophago-gastrectomy

(D) Methods which promote adhesions with a view to the formation of alternative vascular channels

(*a*) Intraperitoneal, omentopexy or the use of talc
(*b*) Intrathoracic, mediastinal packing

It must be remembered that any steps which will divert the flow of portal blood from the varices into other channels may have the effect of opening up these alternative channels to a greater extent, and as it is only the varices in the region of the cardia which are dangerous, any steps which reduce the danger have a good prospect of prolonging the life of the patient, though it must be realised that by closing one outlet for the portal blood we may in fact be raising the portal blood pressure still further There are no records in the literature of long-term results of large series of patients treated by any of the above methods, and therefore it must be assumed that up to date no outstanding success can be claimed for any of them However, some of them may be valuable as emergency measures to control acute haemorrhage from varices and the evidence suggests that most of them carry only slight risk and may do something to prolong life [illegible]

occurred It has mainly been employed as a treatment for cirrhosis of the liver rather than for portal hypertension, i e as a means of relieving ascites That it does give some relief cannot be doubted from the large number of cases reported in the literature but the mortality is high owing to the risk of hepatic necrosis To me it has always appeared to be wrong to deprive a diseased organ of its only arterial blood supply, and although ascites may be temporarily relieved there is no evidence yet that the operation does anything to prolong life or to improve the function of the liver The results are difficult to assess because many patients with ascites are temporarily benefited by a laparotomy only When the operation has been done in many cases the ligature has been placed proximal to the gastroduodenal artery and thus has probably little effect in reducing the arterial blood flow through the liver The enthusiasm for this operation in some quarters has been short-lived and the operation cannot be recommended as a means of treating portal hypertension

The emergency treatment of haemorrhage

It has already been noted that haemorrhage from oesophageal varices [illegible]
therapy Local treatment consists in the use of an inflated balloon to cause pressure on the bleeding point or to obstruct the blood flow T

veins of the cardia and thus cut off the flow to the varices The Sengstaken tube combines both these methods, having one balloon for the oesophagus and another for the stomach A balloon in the oesophagus must be used with caution and at least one example of rupture of the oesophagus by its use has been recorded It seems safer, therefore, if the bleeding does not cease by milder methods, to use a gastric balloon with traction and for this purpose a Miller-Abbott tube will suffice It is not easy to maintain this traction comfortably, if the tube is fixed to the cheek or forehead by adhesive strapping it is likely to cause pain where it presses on the lip, or even cause ulceration in an unconscious patient, while restless patients with neurophathy have been known to pull the tube out even with the balloon inflated Most patients will not tolerate these tubes for more than two or three days, but as a rule this is long enough for the bleeding to stop, if it has not done so by this time more drastic measures should be employed

The tendency has been, as with haematemesis from peptic ulcers, to institute operative treatment earlier and the view is now expressed that if the patient is bleeding so severely that the passage of a tube is necessary an open operation should be undertaken Tubes with inflated bags should only be used as a temporary measure to control bleeding while lost blood is replaced by transfusion and the patient is being prepared for operation

If as a result of previous investigations it is known that the portal vein is patent and the liver function is satisfactory, an emergency portacaval anastomosis may be done if the surgeon is conversant with this operation Alternatively the venogram may be done on the operating table and this is followed immediately by the appropriate operation, if there is no clinical evidence of liver failure and the portal vein is patent, a portacaval anastomosis may be undertaken In such circumstances precautions against the danger of hepatic coma are advisable, blood should be washed out of the stomach and out of the colon, glucose given intravenously, and neomycin given by mouth as soon after the operation as the patient is able to swallow

M C female, aged 46 had been in hospital a year previously with anaemia and ankle swelling, and was found to have a palpable liver and spleen She had a sudden severe haematemesis, followed by a repetition a month later, for which she was transfused with 4 pints of blood and a Sengstaken tube inserted for three days Only six days later she bled again severely and the tube was reinserted and further transfusion given, in the meantime a venogram by intrasplenic injection had shown a patent portal vein and large varices With the tube in place and a blood transfusion running, an ambulance journey of more than a hundred miles was made, and a portacaval

anastomosis was performed next day through a right thoraco-abdominal approach Her gall-bladder which contained stones was removed at the same time The portal pressure had been 42 mm Hg before the anastomosis Convalescence was delayed by a left-sided empyema, but after this had been drained she made a good recovery

In other cases, if the bleeding persists in spite of other measures, a direct operation on the oesophagus must be considered; this is best done through the left pleural cavity In fact, if no surgeon who has experience of portacaval anastomosis is available, this is the best plan, and if a venous shunt is possible this can be done later after there has been time for a fuller investigation of the patient

The question not infrequently arises as to what a surgeon should do who has opened the abdomen at an emergency operation for haematemesis thought to be due to a peptic ulcer, when it is found that there is no ulcer but evidence of portal hypertension In such a case, the steps to be taken must be left to the judgement of the surgeon at the time, they will depend on the patient's general condition, age, and the state of his liver However, if it seems that his haemorrhage is otherwise likely to be fatal, and if there are not too many vascular adhesions, the best plan is to carry out a high gastric transection, with division of the vessels in the lesser omentum and the vasa brevia above the line of section The splenic artery should be divided between ligatures at the same time in order to reduce the blood flowing into the portal circulation At a later date the question of more definitive treatment can be considered

When a decision has been reached that major operation is advisable, it is necessary that the patient should be as fit as possible, preliminary blood transfusion may have been necessary and adequate blood should be available for transfusion at the time of operation The patient should have a diet with an adequate protein content, breathing exercises, with the especial object of obtaining maximum expansion of the bases of the lungs should also be instituted

REFERENCES

BOEREMA, I (1949) Bleeding Varices of the Oesophagus in Cirrhosis of the Liver and Banti's Syndrome" *Arch Chir Neerl*, 1, 253 260

C[illegible] C and F[illegible] P ([illegible]) "N[illegible] Surgical Treatment of Vari [illegible], 27, 422–429

[illegible] Varices by Injection of a Sclerosing Solution *J Amer med Ass*, 135, 754–757

RIENHOFF, W F (1951) "Ligation of the Hepatic and Splenic Arteries in the Treatment of Portal Hypertension" *Bull John's Hopk Hosp*, 88, 368–375

ROUSSELOT, L M (1952) "Autogenous Vein Grafts in Splenorenal Anastomosis a description of techniques and its clinical application in seven patients" *Surgery*, 31, 403–410

SENGSTAKEN, R W and BLAKEMORE, A H (1950) "Balloon Tamponage for the Control of Hemorrhage from Esophageal Varices" *Ann Surg*, 131, 781–789.

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exercised in the selection of the patients Poor selection probably accounts for some of the disappointing results which have been reported but if the indications laid down here are adhered to gratifying results should be obtained The patient should preferably not be above sixty years of age there should be no evidence of recent or rapid deteriora tion in the function of the liver the patient should not be jaundiced and the liver function tests must be adequate Any recent evidence of portal systemic neuropathy should make one very cautious about advising the operation and then it should only be undertaken if the patient is having dangerous haemorrhages As regards the liver function tests the greatest emphasis has been placed on the level of the serum albumin and we have not operated on any patient who had a level below 3 0 gm per cent Re versal of the albumin globulin ratio need not contra indicate the operation if the albumin level is satisfactory We have only used the bromsulpha lein retention test in cases where there has been considerable doubt con cerning the decision but it is generally accepted that retention of more than 30 per cent after 30 minutes indicates that the venous shunt will carry a high risk In two of our patients who had been jaundiced a few months before the operation suggesting at that time that there was active hepatitis the jaundice recurred a few weeks after the operation and per sisted for many months The flocculation tests are not of great value from the point of view of selection for operation but when they are abnormal the risk of post operative neuropathy is greater However many of our patients who have had gro ly disturbed flocculation tests e g a thymol turbidity of 10 units have done well after the operation A raised serum alkaline phosphatase level should not deter the surgeon but if it is high a careful search should be made for any obstruction affecting the extra hepatic bile ducts [illegible]

and the level has remained high after the operation

Technique

Anaesthesia The anaesthesia should be light with the use of a short acting relaxant drug as the right pleura will be opened positive pressure will be needed to maintain expansion of the lung

Position In our experience the only approach which gives an ade quate exposure of the portal vein is by a right thoraco abdominal incision In a thin patient it may be possible to perform the operation from the front but the increased vascularity of the tissues around the common bile duct renders this approach much more d fficult

The patient should be tilted on the left side with pillows under the right shoulder and hip so that the anterior axillary line is uppermost A table

CHAPTER VII

PORTACAVAL ANASTOMOSIS

This operation was first used as an experimental procedure in animals by Eck in Russia in 1877 (for translation—Child, 1953), and first applied to man for the relief of ascites by Vidal (1910) in France Poor results followed these early operations and the method fell into disrepute In

vessel suture, the operation as practised today came to be established The approach through a right thoraco-abdominal incision, as first advocated by Satinsky (1948), was a landmark which has made all the difference to the ease and safety of the operation

Indications

The indications for a portal systemic anastomosis have already been discussed, and it has been concluded that a portacaval anastomosis gives much better long-term results than a splenorenal anastomosis from the point of view of preventing haemorrhage, and carries no greater operative risk Probably there is a greater incidence of neuropathy after this operation, because it is more efficient as a shunt, and as the purpose of the operation is to prevent haemorrhage, the risk of this complication should be taken into consideration when considering whether a shunt should be created Having decided that this is the right line of treatment, the most efficient anastomosis should be performed

Before the operation the patency of the portal vein must be demonstrated This will probably have been done by means of trans splenic venography If the spleen has been removed the best course is to commence the operation by making the anterior part of the incision first, and performing a venogram by injection into a mesenteric vein or one of the veins of the great omentum If the venogram has shown any obstruction of the splenic vein, it is possible that the anastomosis will not drain the splenic part of the portal circulation, and it would be wise to remove the spleen at a preliminary or a second operation

Selection of patients

If good results are to be obtained from the operation, both as regards the immediate mortality and the subsequent course, great care must be

which permits a lateral tilt is an advantage, so that the angle can be adjusted during the operation (Fig 41)

Incision This follows the line of the eighth or ninth rib, and is projected forwards beyond the costal margin for about 5 cm The level of the lower of these ribs should be chosen when the liver is much enlarged A few of the anterior fibres of the latissimus dorsi are divided, the external oblique is split and the rectus sheath opened About 12 cm of the rib are removed subperiosteally, and the corresponding costal cartilage split lengthwise (Fig 42) Further forward the internal oblique and transversalis muscles are incised and the peritoneum opened The pleura is opened through the rib bed and the diaphragm incised radially as far as the bare area, vessels on its surface being secured by stitches, one on each side being left long to hold the diaphragm aside This incision of the diaphragm is important as it allows the liver to be displaced upwards, giving access to its under surface

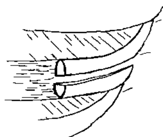


Fig 42 Method of splitting the costal cartilage lengthwise after removal of the rib This allows a more satisfactory repair of the costal margin

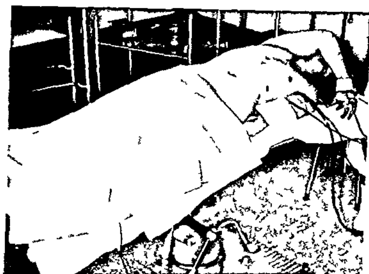
Dissection of the portal vein The next step is usually the most difficult part of the operation, the exposure and dissection of the portal vein The foramen of Winslow is defined (Fig 43) but sometimes it is found to have been obliterated by adhesions An incision in the peritoneum is carried backwards from the foramen along the lower border of the liver in the pouch of Rutherford Morison, this incision should be about 1 cm from the liver so that any vessels, and there may be many collaterals in this fold of peritoneum, can be coagulated or ligatured without damage to the liver The peritoneal incision is carried down the posterior margin of the foramen of Winslow, across its base and up the right free margin of the lesser omentum to the hilum of the liver, a branch incision downwards over the inferior vena cava helps to mobilise the duodenum

In some cases there are enlarged lymph nodes and many thin-walled collateral vessels in the connective tissue along the portal vein, but it is unusual to find any nodes behind the vein, so that this structure is best

but also the gall bladder, the duodenum and adjacent retroperitoneal tissues The common bile duct is retracted forwards and in many cases does not come into view at all Large lymph nodes lying in the line



(a)



(b)

Fig 41 The position of the patient for a portacaval anastomosis The dotted line marks the line of the incision

of approach should be removed During this dissection oozing from tiny vessels may be troublesome, but it is best controlled by patient pressure and the cautious use of diathermy On more than one occasion we have inserted a pack and adjourned for refreshment at this stage of the operation, but the knowledge that a patent portal vein has been shown by venography is an encouragement to continue with patient dissection

When once the portal vein is exposed it is found to lie in a tunnel of connective tissue, to which it is only loosely adherent, and it can be dissected free using small gauze pledglets (Fig 44) Careful watch must be kept for any tributaries as troublesome bleeding will occur if they are torn, they should be divided between ligatures of the finest silk These tributaries are inconstant, but one is not infrequently found entering the left lateral wall of the vein, i e the side away from the operator

When there has been some thrombosis in the vein adhesions are found outside the vein to the adjacent connective tissue which make the dissection much more difficult, previous venography will have demonstrated that there is a fairly wide lumen, so in the cases suitable for a portacaval anastomosis any thrombus present is likely to be attached to the wall on one side of the vein only Sometimes mural thrombi can be successfully removed from the vein after it has been divided during the next stage of the operation but if the lumen is adequate and the thrombi are firmly adherent, they should not be disturbed

As soon as a ligature can be passed round the vein the latter can be drawn aside which facilitates further dissection upwards and downwards The vein should be freed down to the upper border of the duodenum and upwards to the hilum of the liver, preferably so that its division into the right and left branches can be defined Ligatures are then placed round these branches but not tied (Fig 45) In some cases the division of the vein is very high and, provided an adequate length of the main vein is freed it is not necessary to expose the branches

Exposure of the vena cava Only the anterior surface of this structure need be exposed, and the adventitia should be removed over an area of about 5 cm by 3 cm This means that it is cleared from the level of the entrance of the left renal vein up to its disappearance behind the liver Occasionally one or two small tributaries require to be ligatured and divided, but it is important to make the anastomosis as high as possible in the vena cava This is a convenient time in the operation to measure the portal venous pressure, either by a canula in a tributary or a needle in the portal vein itself (Fig 6)

Making the anastomosis The next step is the division of the portal vein The ligature round the upper end of the portal vein, or those on its two branches, are tied and a Blalock clamp placed on the main vein as low down as possible, there should be at least 1 5 cm of the vein between the clamp and the point of division (Fig 46) Blood is aspirated from the



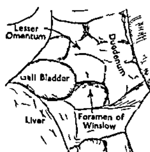


Fig 43 Portacaval anastomosis Exposure of the right free border of the lesser omentum and the foramen of Winslow



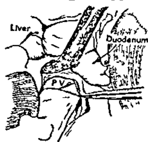


Fig 44 Portacaval anastomosis The portal vein exposed in the margin of the lesser omentum

vein to ensure that the clamp is sufficiently tight before the vein is divided, but if it is suspected that there is some clot in the vein it is better to cut the vein across before the clamp is tightened so that any loose clot may be washed out

The vein is now cleanly cut across just below the ligatures and washed clear of blood If there is any recent clot present it is wise to release the clamp to ensure that there is an adequate flow of blood Before doing this two stay stitches are placed in the cut end of the vein, so that its retraction behind the duodenum is prevented, the end of the vein is then compressed between the left thumb and index finger while the clamp is removed On releasing the grasp it will be seen if there is a good flow of blood, and if so, the Blalock clamp is reapplied while the end of the vein is again compressed between the thumb and index finger If there is not a good flow, clot may be removed by a strong sucker, or by gall-stone forceps passed along the vein (Fig 47) It is important to ensure that there is a wide lumen before the clamp is reapplied If clot has been present 1 ml (5000 units) of heparin should be injected into the vein behind the clamp

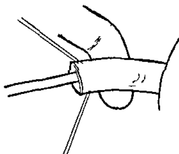


Fig 47 Portacaval anastomosis Method of removing blood clot from the portal vein

A suitable clamp (such as the Satinsky clamp) is now placed so that it picks up part of the anterior wall of the vena cava, and this is also aspirated to ensure that the clamp has efficient control (Fig 48) A vertical incision is made in the vena cava, its length being half the circumference of the portal vein (Fig 49) The anastomosis is made with arterial sutures using a continuous everting stitch, this being less likely to initiate thrombosis than a simple over and over stitch It is helpful to put a stay stitch in the outer edge of the incision in the inferior vena cava to hold it aside while the posterior suture line is being inserted The medial wall is sutured first using a continuous everting mattress stitch and starting at the top and when the suture has been pulled tight it is secured at the lower end by a lock stitch (Fig 50) A fresh stitch for the lateral wall is tied to the upper end of the medial wall suture, and when the suturing is completed it is secured to the lock stitch at the lower end As soon as the suturing is completed the clamps are removed when as a rule there is hardly any leakage, light pressure on the suture line may be necessary for a few minutes, but it is unusual for any additional stitches to be required In many cases it is possible to see through the thin wall of the vena cava the turbulence of blood caused by the portal inflow (Fig 51)



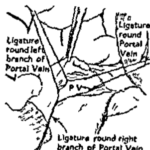


Fig 45 Portacaval anastomosis The portal vein has been freed, and ligatures have been placed round it, and round its two main divisions



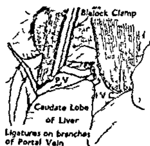


Fig 46 Portacaval anastomosis A Blalock clamp has been placed on the portal vein and the ligatures tied The vein has been aspirated to ensure that the clamp and ligatures are tight

If desired a second reading of the portal pressure may be taken at this stage and before the wound is closed a small portion of liver should be taken for histological examination This should be from the surface of the right lobe well away from the edge or from the gall bladder, in order to obtain a sample which is representative of the liver as a whole

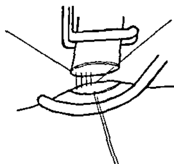


Fig 50 Portacaval anastomosis The posterior layer of the suture using a continuous mattress stitch

Closure Drainage of the pleura is unnecessary, but a tube should be inserted into the pouch of Rutherford Morison and brought out through the anterior end of the wound and led to an underwater seal It can usually be removed on the following day The diaphragm is repaired with interrupted stitches and the pleura closed by suture of the periosteum of the rib bed The remainder of the incision is closed in layers

A blood transfusion will have been required during the operation in



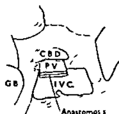


Fig 51 Portacaval anastomosis The completed anastomosis the portal vein is seen emerging from behind the common bile duct



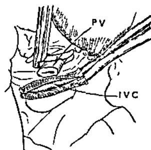


Fig 48 Portacaval anastomosis The portal vein has been divided and a lateral clamp picks up a portion of the anterior wall of the vena cava



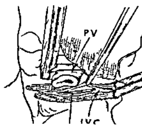


Fig 49 Portacaval anastomosis Incision in the wall of the vena cava

CHAPTER VIII

SPLENORENAL ANASTOMOSIS

This was the first type of venous shunt to be employed during the period of the revival of interest in vascular surgery which has taken place during the last fifteen years or so, for Blakemore and Lord's first cases were treated by removal of the spleen and the left kidney (1945), and the end-to-end anastomosis of their respective veins It was not long before it was realised that there was no need to sacrifice a healthy kidney and that an anastomosis to the side of the renal vein was as effective In our experience, however, it has not been such a satisfactory procedure as a portacaval anastomosis, we have only employed it in those cases, mostly children with extra-hepatic obstruction, in which the portal vein has not been suitable The operation itself involves the removal of the spleen, no small item when there are vascular adhesions, the vein is not so satisfactory as the portal vein for holding sutures and, especially in children, it may not be large enough for a satisfactory opening On the whole, therefore, it is a more formidable operation than a portacaval anastomosis, and should be reserved for those patients in whom the latter is not possible, in whom the liver function is adequate (the same indications apply in this respect as for a portacaval anastomosis, see Chapter VII), and in whom the splenic vein is large It will be seen then that the numbers requiring this operation are few, and in fact during the last seven years we have only performed it ten times compared with eighty-five portacaval anastomoses

The operation is best carried out through a left abdomino-thoracic incision along the bed of the ninth or tenth rib The patient is supported with the anterior axillary line uppermost It is a wise precaution to open the lesser sac through the gastro-splenic ligament early in the operation (Fig 52), and dissect out and ligature the splenic artery before the spleen is handled This reduces the vascularity of the peritoneal folds which carry collateral vessels There are two special points in the removal of the spleen, the first is that much oozing may be avoided if the vessels in the lieno-renal fold of the peritoneum and those passing up from the spleen towards the diaphragm are coagulated by diathermy before they are divided (Fig 53), the second is that the splenic vein must be preserved and handled very gently. The vein is kept intact until all the other attachments of the spleen have been divided, it is then secured by a bull-dog clamp and divided, and any blood distal to the clamp is washed out Between 3 and 5 cm of the vein should be freed, and this may entail

most cases, in order to replace any blood loss During the post-operative period a watch must be kept for a pleural effusion which may require aspiration The most serious complication is the neuropathy which may be precipitated by the operation, the signs and treatment of this are discussed in a later chapter It has not been the custom to administer anti coagulants during the post-operative period and their use is regarded as unnecessary

REFERENCES

[illegible] A H and Lord J W (1945) "The Technique of Using Vital- [illegible] ical Shunts for Portal Hypertension " [illegible] " *Surg Gynec Obstet*, 96 375-376

Eck, N V (1877) "Concerning Ligation of the Vena Porta" *Voen med J St Petersburg*, 130, 1-2

Satinsky, V P (1948) "Thoraco abdominal Approach for Portacaval Anastomosis" *Ann Surg*, 128, 938-947

Vidal, E (1910) "Discussion of the Eck Operation" *Rev Chir, Paris*, 42 1181-1182

CHAPTER VIII

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Fig 52 Splenorenal anastomosis Exposure of large collateral vessels in the gastro splenic ligament



Fig 53 Splenorenal anastomosis Exposure of the lieno renal fold of peritoneum by drawing the spleen forwards

ligature and division of a few small tributaries coming from the tail of the pancreas

The next step is the mobilisation of the kidney and the preparation of its vein An incision is made in the peri-renal fascia and the kidney delivered from its bed; the vessels of the hilum are next dissected out, and to do this it is helpful to place a tape under the hilum and over the upper and lower poles so that traction on the tape draws the hilum of the kidney up *into the wound (Fig 54) Both the artery and the vein should be* dissected out as, if the artery is not occluded while the anastomosis is being



Fig 54 Splenorenal anastomosis Drawing the kidney forwards by means of a tape passed behind the hilum

made, the kidney becomes very engorged At least 3 cm of the vein require to be isolated, which may entail ligature of one or two small tributaries Some surgeons use a small clamp which only occludes part of the lumen, but owing to the size of the vein we have found it simpler to occlude the vein completely The actual anastomosis does not as a rule take longer than twenty minutes, and we have seen no evidence of damage to renal function as shown by post-operative intravenous pyelography, when the renal function has been controlled for as long as this However, if more time *is required it is advisable after twenty minutes to release the clamp* on the renal artery for a few seconds and thus allow some fresh arterial blood to flow into the kidney

When all is ready for the actual anastomosis, an encircling ligature is placed round the renal vein close to the kidney or preferably one round

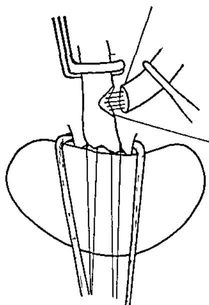


Fig 55 Splenorenal anastomosis Suturing the end of the splenic vein to the side of the renal vein by a continuous everting stitch



Fig 56 Splenorenal anastomosis The completed anastomosis.

each of its two main tributaries, and traction of this ligature occludes the vein A Blalock clamp is very suitable for the proximal end of the vein, as it allows the assistant to hold the vein steady Before these are tightened the bull dog clamp is placed on the renal artery A transverse incision is made in the upper wall of the vein, the splenic vein brought down to it and the anastomosis is made with a continuous everting stitch (Fig 55) If the splenic vein is small it should be cut obliquely to increase the size of the lumen of the anastomosis When the anastomosis is complete the clamps are removed and the kidney returned to its bed, making sure that there is no kinking of or pressure on the splenic vein (Fig 56) The wound is closed with a drain down to the region of the anastomosis for the first twenty-four hours

The post operative management of this operation is the same as that after a portacaval anastomosis

REFERENCE

BLAKEMORE A H and LORD J W (1945) The Technique of Using Vitallium Tubes in Establishing Portacaval Shunts for Portal Hypertension' *Ann Surg*, **122**, 476–489

CHAPTER IX

DIRECT OPERATIONS ON THE VARICES

1 *Injection therapy* This was introduced by Crafoord and Frenckner (1939) who used a sclerosing solution injected through a needle passed down an oesophagoscope When the size of these vessels is noted at an operation it must be realised that the prospects of getting adequate thrombosis by this method are remote, the method has been used very little during recent years Macbeth (1955) reported his results in 30 patients but when the difficulties of assessing the prognosis are considered it is not easy to judge the value of this method Repeated passage of the oesophagoscope is necessary, his patients having injections on the average on five separate occasions He advocates removal of the spleen as an adjunct to injection therapy

2 *Portal-azygos disconnection or gastric transection* Tanner (1950) described an operation which was intended to divert the portal blood from the dangerous area of the oesophageal varices, this consisted of dividing all the venous channels in the wall of the body of the stomach, the lesser omentum and the gastro-splenic ligament above the level of the line of section of the stomach, which he divided across completely and resutured (Fig 57) Later Tanner (1958) added the freeing of the lower 5 cm of the oesophagus and upper 5 cm of the stomach of all their vascular connections, for injection specimens had shown that blood was still reaching the varices by veins around the cardia, this revised operation has been done by him in 11 cases which were bleeding at the time with early control in every case In theory this is a good operation, but it is difficult to perform, particularly if there are vascular adhesions to the stomach and spleen, and the venous channels tend to reform across the line of suture In order to prevent this Meredino and Dillard (1955) have suggested the interposition of a segment of jejunum between the stomach and oesophagus, but as the pressure in the jejunal veins is high it hardly seems that the prospects of fresh anastomoses would be lessened by this procedure

3 *Oesophageal transection* In view of the technical difficulties sometimes encountered in making a portal-azygos disconnection by the abdominal route, I have carried out a number of similar operations through the left pleural cavity, transecting either the lower end of the oesophagus or the fundus of the stomach after incision of the diaphragm When transecting the stomach by this route care is taken at the same time to divide the upper ends of the lesser omentum and the gastro-splenic liga-

ment as in the operation devised by Tanner In the majority of cases, however it is the oesophagus which has been transected This is a simple and safe operation except when operating as an emergency for haemorrhage It may be said that the veins are divided too high up above the point where rupture occurs This is a valid argument, but in fact the results of oesophageal transection have been no more unsatisfactory, in fact a little better than operations on the stomach Though fatal haemorrhage does occasionally *occur from varices in the fundus of the stomach, this is rare when* compared to the risk of bleeding from the oesophagus It is probable that

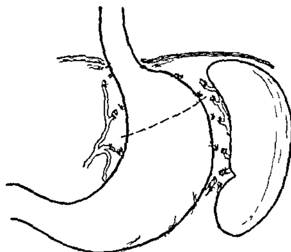


Fig 57 *Gastric transection with removal of the spleen* The transection is made about 5 cm below the cardia and all vessels to the upper third of the stomach are divided

some of the varices below the line of section become thrombosed as a result of stasis of their contained blood after the section In the earlier cases in the series the oesophagus was divided completely across just above the diaphragm and resutured, but after a time it was appreciated that nearly all the varicose channels lie in the mucosa and sub-mucosa, and it was very unusual to find any in the muscular coats There was therefore no point in dividing the muscle, so that in many of the cases the muscular coat has been split lengthwise, and only the mucosa and sub-mucosa divided horizontally and resutured (Fig 58) At the same time any large vessels on the outer surface of the lower oesophagus are ligatured

The technique is as follows The left pleura is opened through the

rib-bed of the eighth or ninth rib, the pleura over the oesophagus incised, and the lower 5 cm of the oesophagus mobilised and lifted from its bed by tapes A light clamp is then placed across the lower end immediately above the diaphragm, the muscle incised longitudinally for about 5 cm and the mucosa, which separates easily from the muscular coat, is lifted out, cut across transversely and resutured by a continuous catgut stitch The clamp is then removed and any bleeding points at the lower end are underrun by catgut stitches A few sutures bring the muscular fibres together and any large veins on the surface below the level of the section are also underrun by sutures The oesophagus is then replaced in the mediastinum, with a few stitches in the mediastinal pleura, and the chest

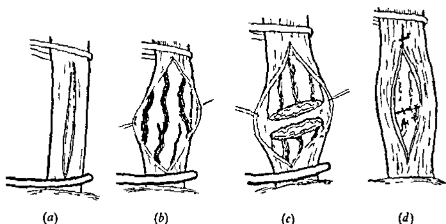


Fig 58 Oesophageal transection (a) The muscle is split for about 5 cm (b) The mucosa is exposed (c) The mucosa and submucosa are cut across (d) The mucosa and submucosa are resutured with a continuous catgut stitch.

closed, with or without a temporary drain, depending on whether all oozing has been completely stopped

When a gastric transection is carried out through the thorax, a radial incision is made in the diaphragm leaving the fibres around the oesophageal hiatus The fundus of the stomach is then drawn up and divided between clamps a little below the cardia After division of the upper short gastric vessels and the upper end of the lesser omentum, the stomach is resutured in two layers and then the diaphragm and the chest wall closed During the last few years this operation has, with one exception, been performed only in children in whom it was thought that oesophageal transection might lead to some stricture formation

Oesophageal or gastric transection has been advocated under three circumstances in patients who have had serious bleeding first when there

was no vein suitable for an anastomosis (Fig 59) second if the liver function was considered too poor for an anastomosis or third in patients who had had evidence of portal systemic neuropathy It may also be advised in patients who are above the age level at which a venous shunt can be recommended

4 *Ligature of the varices* Boerema (1949) described as an emergency operation incision of the oesophagus lengthways by a left transpleural



Fig 59 Trans splenic venogram in a patient with extra hepatic obstruction although the splenic vein was patent it was considered too small for a splenorenal anastomosis and an oeso phageal transection was done Note the collateral circulation fills the left subcostal vein

venous anastomosis after only a short interval

5 *Mediastinal packing* This operation was suggested by Garlock and Som (1950) as a means of promoting vascular adhesions on the outside of the oesophagus which might drain the blood away from the varices inside

and thus reduce the pressure inside them. The upper mediastinum was approached through an incision in the neck and the lower oesophagus by a right pleural approach; gauze strips were placed alongside the oesophagus and their ends left protruding through the wound or through a separate stab incision; they were removed after about fourteen days.

6. *Oesophago-gastrectomy.* Excision of the varicose bearing area was first recommended by Phemister and Humphreys (1947) and has since been employed by Macpherson and others (1956). It would be thought that new venous channels are just as likely to develop across the new suture line after this operation as after a gastric or oesophageal transection. There are very few patients who have had a resection of the fundus of the stomach and cardiac sphincter for other lesions, especially for malignant growths, who are able to enjoy good digestion afterwards. The risk of such indigestion is quite a high price to pay for the benefits of the operation, which are problematical, and in a condition in which the prognosis is so unpredictable. So far only a few patients treated by this rather drastic method have been recorded and no results are available which would indicate that it offers a greater freedom from recurrence of haemorrhage than other operations which carry less operative risk.

7. *Promotion of intra-peritoneal adhesions.* Rutherford Morison of Newcastle and Talma of Utrecht independently introduced the operation of omentopexy and scarification of the liver with a view to the promotion of adhesions which would provide portal systemic anastomoses. Their operations were done as a means of treating ascites, particularly associated with alcoholic cirrhosis, and met with some success, especially if the patients abstained from the taking of alcohol. Grey Turner reported some patients who had survived for many years after this operation, but the fickle prognosis of the disease makes the assessment difficult. As a means of preventing haemorrhage there does not seem to be any place for this type of operation, but where there is evidence that the main cause of the ascites is due to obstruction to the outflow tract from the liver, and other methods fail to give relief, there may be a justifiable reason for attempting to promote vascular adhesions between the liver and the parietal peritoneum. Talc has been employed for this purpose in 2 of my own cases and its use has also been reported by Madden and others (1954), but there are not sufficient cases so far described to judge its long term effect on the course of the disease.

REFERENCES

BOEREMA, I. (1949). 'Bleeding Varices of the Oesophagus in Cirrhosis of the Liver and Banti's Syndrome.' *Arch. Chir. Neerland.*, 1, 253–260.

CRAFOORD, C. and FRENCKNER, P. (1939). "New Surgical Treatment of Varicose Veins of Oesophagus." *Acta Otolaryng.*, 27, 422–429.

CRILE G (1957) Transesophageal Ligation of Bleeding Varices *Surgery* **42** 583 584

GARLOCK J H and SOM M L (1950) Further Observations on Packing of Mediastinum for Esophageal Varices *J Thorac Surg* **19** 572 586

LINTON R R and ELLIS D S (1956) Emergency and Definitive Treatment of Bleeding Esophageal Varices *J Amer med Ass* **160** 1017 1023

MACBETH R (1955) Treatment of Oesophageal Varices in Portal Hypertension by means of Sclerosing Injections *Brit med J* **2** 877 880

MACPHERSON A I S OWEN J A and INNES J (1956) Surgical Treatment of Portal Hypertension *Lancet* **1** 353 357

MADDEN J L LORE J M GEROLD F P and RAVID J M (1954) The Pathogenesis of Ascites and a Consideration of its Treatment *Surg Gynec Obstet* **99** 385 391

MEREDINO K A and DILLARD D H (1955) The Interposed Jejunal Segment at the Oesophago gastric Junction *Ann Surg* **142** 486 509

HEMISTER D B and HUMPHREYS E M (1947) Gastro esophageal Resection and Total Gastrectomy in the Treatment of Bleeding Varicose Veins in Banti s Syndrome *Ann Surg* **126** 397 410

TANNER N C (1950) Discussion on Gastroduodenal Haemorrhage as a Surgical Emergency *Proc R Soc Med* **43** 147 152

—— (1958) Operative Management of Haematemesis and Melaena *Ann R Coll Surg* **22** 30–42

CHAPTER X

PORTAL SYSTEMIC NEUROPATHY

It is only in the last few years that it has been shown that some of the neurological symptoms which are found in cases of liver disease are due to the shunting *of portal blood direct into the systemic circulation*, whence it reaches the brain before it has passed through the liver (Sherlock *et al*, 1954) As the treatment of portal hypertension which is advocated includes methods which increase the volume of blood which by-passes the liver it is only to be expected that these *neurological effects may follow* such treatment (McDermott *et al* 1954) Although these manifestations do in fact occur in many patients who have had no operation, and are due to the natural collaterals which have developed, it has been found that they are the most serious complication of the shunt operations It follows therefore that the surgeon must consider this risk in each particular case if operation is advised, and must be able to treat the complication if it arises during the post-operative period It thus occurs that the more efficient the venous shunt in diverting blood from the dangerous varices, the greater is the risk of neurological complications There can be no doubt that an end-to-side portacaval anastomosis, which diverts all the portal blood from the liver, is much the most efficient as a means of preventing haemorrhage from the alimentary tract in cases of portal hypertension, but it carries a greater risk of neuropathy than the less efficient splenorenal anastomosis The difference is such that some writers have advocated a splenorenal anastomosis if the splenic vein is large, in preference to the portal vein-vena cava type of shunt With this argument I cannot agree, for if the patient's life is endangered by haemorrhage, the most efficient operation which will prevent that haemorrhage should be employed, if the patient has had evidence of neuropathy before operation, then it is probably unwise to add in any way to the venous shunt and an alternative operation on the varices would be indicated Patients whose liver function is poor have a greater tendency to this type of neuropathy, and this is an argument in favour of limiting shunt operations to patients whose liver function has not deteriorated too far, as laid down in the indications for the operations in Chapter VII

The precise nature of the toxic substance or substances which cause these effects on the brain has not yet been determined In some respects the symptoms resemble those of ammonia intoxication and many of the patients have raised blood ammonia levels, but this has not been a con

stant finding; there is no doubt that some protein breakdown products containing nitrogen are responsible, but further work is required to identify them and to find a specific neutralising agent. It has been shown that there may be a rise in the level of certain keto acids in the blood (Dawson *et al.*, 1957) but there is no proof that they are the responsible substances. Modern therapy, which has met with considerable success, is aimed at preventing such substances gaining access to the circulation rom the alimentary canal.

Though this concept of portal systemic neuropathy has been generally cepted, patients with liver failure may have serious disturbances of nsciousness, even when there is little or no collateral circulation and this may in fact be due to lack of substances in the blood stream which are normally supplied only by the liver, and which are essential for central nerve function. It has been suggested that 5-hydroxytryptophane may be such a substance (Bessman, Merlis and Borges, 1957).

The symptoms

The neuropathy manifests itself in a variety of ways but in the same patient each episode tends to hold to a constant pattern. The most usual symptom is increasing drowsiness passing to coma, but some patients may pass very suddenly from an apparently normal state to complete unconsciousness, and equally suddenly revert to their fully alert condition. In the mild cases the patients have expressionless features, are slow in answering questions and may not always give rational answers. It is at this time that the characteristic flapping tremor of the hands may be elicited if the patient is asked to hold them out in front of him. Other patients may be quite conscious but irrational and say and do peculiar things, as a result they may be considered psychotic cases and be sent to a mental hospital. An unusual manifestation is excessive salivation; one of our patients suffered severely from this for two years after a portacaval anastomosis had been performed, though in every other way he behaved quite normally. In a few instances there may be personality changes, and though the patients may seem quite normal to those who do not know them well, their relatives say that there has been some change in their behaviour, which the patient does not recognise himself.

The syndrome is much more likely to occur in elderly patients, thus in one series recorded (Summerskill *et al.*, 1956) among 17 patients there were only 2 under the age of 40 years, and the average age was 49. In 4 of these 17 cases a portacaval anastomosis had been performed, in the remainder there was a natural collateral circulation. In the writer's series of portacaval anastomoses the average age of 80 patients was 31 years, but among the 16 who have shown signs of neuropathy after the operation the average age was 42.5 years.

According to Summerskill *et al* (1956) the blood ammonia level is usually raised and in the cerebro-spinal fluid increase in both the ammonia and glutamine levels are constant findings The electroencephalogram shows slowing of the dominant rhythms and irregular wave patterns

Factors which precipitate an attack are either a large protein meal or a gastro oesophageal haemorrhage which has the same effect in introducing protein into the alimentary canal Some patients can tolerate a low protein diet but show symptoms when this is raised above a certain level Their tolerance of protein may not remain constant and probably depends on variations in their liver function

In the first 80 patients of our series of portacaval anastomoses neurological complications have been noted in 8 cases during the early post-operative period, all 4 patients who have died while still in hospital have succumbed from this cause, it is possible that with our present knowledge of the ætiology and the treatment available, some of them might have been saved

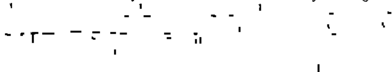
Two patients, aged 38 and 31, became comatose on the day following operation, the older patient never fully regained consciousness and died on the thirty-first post-operative day, no definite cerebral changes being found at autopsy The younger patient who also had bronchiectasis, re-
... coma on the fifth day and was making good progress until

died four days later The third fatal case, a man of 45 years who had extensive thrombosis in his portal system had a small haemorrhage on the fifth post operative day and relapsed into coma, dying two days later The fourth a woman of 57 years showed no abnormal signs for four days but then became drowsy and remained in varying states of consciousness until she died in coma six weeks after her operation In one case there was a bout of coma which lasted from the third to the fifth post operative day and the patient has shown no sign of any recurrence, another case had intermittent coma for three months, but then fully recovered and died of an unrelated cause (ruptured aortic aneurysm) two years later An example of the danger of blood in the intestine is given by a man who ... years after his portacaval anastomosis underwent a gastrectomy

operation Ever since, he has had to restrict his protein ...
liable to get ' giddy and fainting" turns Eight patients after portacaval anastomosis have had episodes of neuropathy after they returned home, in two, severe attacks leading to coma have happened within a few days of leaving hospital, and patients should be warned against celebrating the occasion by a large protein meal, in neither case has there been any recurrence, though they have both been followed for two years

Treatment

As some of the patients who have had portal systemic neuropathy have had normal liver function before operation and have been able to take a high protein diet caution should be exercised in the early post operative course of all patients who have had a shunt operation They should start with a very low protein diet but if all is well this can be increased by about 5 gm protein a day until they are on a normal diet If any signs of drowsiness or unusual mental behaviour appear a protein free diet should be at once reverted to and if the symptoms disappear the increase may be tried again If the state of consciousness deteriorates more active measures must be instituted The colon should be washed out and an enema given in order to remove the protein in the alimentary canal as quickly as possible if there is a possibility that haemorrhage into the stomach has occurred a stomach tube should be passed and the gastric contents aspirated The patient should also be given an antibiotic in order to abolish the bacterial activity in the intestine at first aureomycin was given



may help to prevent pulmonary complications and in the unconscious patient it may be necessary to aspirate secretions from the trachea Intravenous fluids may be required in order to maintain fluid balance Walshe (1956) has recommended the use of intravenous sodium glutamate 20 gm in a litre of saline on the basis that this substance removes ammonia by converting it into glutamine we have used it on a number of occasions but are unable to convince ourselves that it has any specific action

There are some patients who find themselves unable to take a diet containing a normal amount of protein and as a result of restriction of protein intake feel some lassitude and are unable to do their work such patients may be able to tolerate a fuller diet if they are kept on a small maintenance dose of aureomycin or neomycin and it is worth giving this a trial

When advocating a shunt operation the risk of portal systemic neuropathy should always be kept in mind in some patients who have had severe haemorrhage and for whom a shunt is likely to be the only way of preventing a recurrence this risk will have to be taken in others the risk may seem too great At the moment this complication provides the greatest anxiety concerning this method of treatment of portal hypertension

REFERENCES

[illegible] "Effect of 5-Hydroxy- [illegible] Coma" *Proc Soc* [illegible]

DAW [illegible]

DAW [illegible]

McDERMOTT, W V, ADAMS, R D and RIDDELL, A G (1954) "Ammonia Metabolism in Man" *Ann Surg*, 140, 539–554

SHERLOCK, S, SUMMERSKILL, W H J, WHITE, L P and PHEAR, E A (1954) "Portal-Systematic Encephalography Neurological Complications of Liver Disease" *Lancet*, 2, 453–457

SUMMERSKILL, W H J, DAVIDSON, E A, SHERLOCK, S and STEINER, R E (1956) "The Neuropsychiatric Syndrome associated with Hepatic Cirrhosis and an Extensive Portal Collateral Circulation" *Quart J Med*, 25, 245–266

WALSHE, J M (1956) "Hepatic Coma" *Post Grad med J*, 32, 467–472

CHAPTER XI

PORTAL HYPERTENSION IN CHILDREN

Portal hypertension presents a number of special problems when it occurs in children so that the subject deserves a chapter to itself It has already been mentioned that it is the most common cause of haematemesis in this age group and any child who has this symptom must be fully investigated with this in mind It is found in practice that in the great majority of all cases of portal hypertension the lesion starts as an intra hepatic obstruction but to this children are an exception owing partly to the relative rarity of

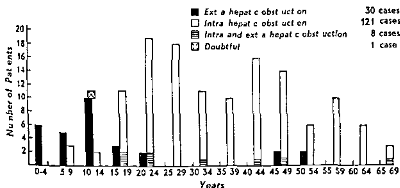


Fig 60 The age at which haematemesis first occurred in 160 patients There is a preponderance of cases of extra hepatic obstruction below the age of 15 years

cirrhosis of the liver at this age The other most important factor is the frequency of thrombosis of the portal vein in children giving rise to extra hepatic obstruction There are a number of cases on record in which there has been a definite history of umbilical infection and it is presumed that this has led to a septic phlebitis of the umbilical and portal veins, but in many cases no such history can be obtained yet in my series it is not until the fourth quinquennium of life is reached that the number of cases of haemorrhage due to intra hepatic obstruction exceeds those due to obstruction of the portal vein outside the liver As a result of this dis tinction treatment is a much greater problem in children in spite of the fact that they have no disease of the liver

Figure 60 shows the age at which the first haemorrhage occurred in 160

REFERENCES

BESSMAN, S P, MERLIS, J K and BORGES, F (1957) "Effect of 5-Hydroxytryptophane upon Electroencephalogram in Hepatic Coma" *Proc Soc exp Biol*, 95, 502–504

DAWSON, A M, de GROOTE, J, ROSENTHAL, W. S and SHERLOCK, S (1957) "Blood Pyruvic Acid and Alpha-ketoglutaric Acid Levels in Liver Disease and Hepatic Coma" *Lancet*, 1, 392–396

DAWSON, A M, McLAREN, J and SHERLOCK, S (1957) "Neomycin in the Treatment of Hepatic Coma" *Lancet*, 2, 1263–1268

McDERMOTT, W V, ADAMS, R D and RIDDELL, A G (1954) "Ammonia Metabolism in Man" *Ann Surg*, 140, 539–554

SHERLOCK, S, SUMMERSKILL, W H J, WHITE, L P and PHEAR, E A (1954) "Portal-Systematic Encephalography Neurological Complications of Liver Disease" *Lancet*, 2, 453–457

SUMMERSKILL, W H J, DAVIDSON, E A, SHERLOCK, S and STEINER, R E (1956) "The Neuropsychiatric Syndrome associated with Hepatic Cirrhosis and an Extensive Portal Collateral Circulation" *Quart J Med*, 25, 245–266

WALSHE, J M (1956) 'Hepatic Coma" *Post Grad med J*, 32, 467–472

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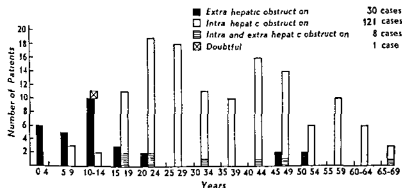


Fig 60 The age at which haematemesis first occurred in 160 patients. There is a preponderance of cases of extra hepatic obstruction below the age of 15 years

cirrhosis of the liver at this age. The other most important factor is the frequency of thrombosis of the portal vein in children giving rise to extra-hepatic obstruction. There are a number of cases on record in which there has been a definite history of umbilical infection and it is presumed that this has led to a septic phlebitis of the umbilical and portal veins, but in many cases no such history can be obtained, yet in my series it is not until the fourth *quinquennium of life is reached that the number of cases of* haemorrhage due to intra-hepatic obstruction exceeds those due to obstruction of the portal vein outside the liver. As a result of this distinction, treatment is a much greater problem in children in spite of the fact that they have no disease of the liver.

Figure 60 shows the age at which the first haemorrhage occurred in 160

patients who bled from oesophageal or gastric varices, this group is not representative of bleeding in cases of portal hypertension as a whole for many of these patients had been selected as suitable for surgical treatment before they were referred to me, probably many in the older age groups were excluded, but the purpose is to show the relative distribution of the intra- and the extra-hepatic groups It will be noted that all except four of the extra-hepatic cases had their first haemorrhage before the age of 25 and that there was no case of intra-hepatic obstruction who had his first bleeding before the age of 5, though more recently a case of congenital fibrosis of the liver has been seen who bled for the first time at the age of 3

From what has been said it is clear that most cases of portal hypertension in children are due to thrombosis of the portal vein, that they have normally functioning livers and that haemorrhage is the only symptom If untreated most of these children will eventually die of haemorrhage, but if this can be prevented they will lead perfectly normal lives This is the tragedy, for very often these little patients have no suitable vein in the portal system with which a wide anastomosis can be made to a systemic vein, and thus there is no possibility of bringing the pressure in the portal system down to normal The portal vein is cavernous and replaced by many collateral channels of varying size, but none of them suitable for an anastomosis either because they are individually too small or because their walls are thin and tear easily In some of these cases the splenic vein is also thrombosed and in the smaller children it is not large enough for an anastomosis which has a good chance of remaining patent One therefore has to turn to the less effective operative procedures on the oesophagus or stomach to prevent these recurrent haemorrhages.

Jordan and his colleagues (1956) have reported the results of 9 children treated by various methods, 3 had cirrhosis of the liver and in 1 the portal vein was also cavernous In 2 patients in whom a dilated varix around the obliterated portal vein was used for an anastomosis to the vena cava, the result was unsatisfactory When the splenic vein was patent, a splenorenal anastomosis done in three patients gave good results, in a 17-year-old patient in whom bleeding persisted after splenectomy, resort was had to a total gastrectomy, but the follow-up was only over a period of nine months These writers condemn splenectomy alone and consider that, even if bleeding has not occurred, a splenorenal anastomosis should be performed, but they regard 4 years as the lowest age for this owing t the small size of the splenic vein in infants They point out, however, that in some cases of portal vein stricture, this vein may be suitable for a portacaval anastomosis

Ekman (1957) reports a very similar experience In 30 patients with extra-hepatic obstruction in whom anastomoses were made, 22 were under the age of 20 when they had their first haemorrhages Eighteen of these had splenorenal shunts and 4 had makeshift shunt operations One of the

last group died after the operation and the other 3 have all bled again Amongst the 18 patients with splenorenal anastomoses, 9 out of the 16 survivors have had recurrent haemorrhage, and 4 have succumbed

Shumaker and King (1952) in a study of 21 patients who started having bleeding from varices before the age of 15 found that in only 1 was there evidence to suggest intra-hepatic obstruction Five of these children had no operative treatment and 4 of them died of haemorrhage within a few months of their first attack The only survivor did not have her first bleeding until the age of 14 and further attacks continued at frequent intervals for the next 7 years In 10 patients splenectomy alone was the first operation performed and only 1 in whom the operation was not carried out until she was 32 years old, had not had further bleeding at the time of the report, this again points to the inadequacy of splenectomy as a means of treatment Oesophago-gastrectomy was done in 5 patients with 2 operative or post-operative deaths, all the cases had had previous splenectomy, and 2 of the survivors had no further bleeding when followed for three and a half and two years respectively Bleeding recurred within one and a half years in 2 cases who had devascularisation of the oesophagus and stomach combined with vagotomy and gastrojejunostomy Eight anastomosis operations were carried out on 7 patients, 4 with splenorenal shunts had done well but in 2 others bleeding recurred, the other 2, in which the portal vein and a mesenteric vein were used for anastomosis, had only been followed for a short time

The disappointing results of direct operations on the stomach or oesophagus have led Koop and Roddy (1958) to try inserting a piece of transplanted colon, retaining its blood supply between the oesophagus and stomach after partial oesophago-gastrectomy, they have done this operation in 5 children, but in none was the follow-up longer than a year at the time of their report

In my own series there are 45 patients who had their first haemorrhage from oesophageal varices before they reached the age of 20 In 19 the obstruction was intra-hepatic and the age of onset is noted in the following table

Cause of cirrhosis where known	*No of cases*	*Age of onset of haemorrhage*
No cause found	10	11, 12, 15, 16, 17, 18, 19, 19 19, 19
Congenital fibrosis of the liver	4	3 5 6, 17
Congenital absence of bile ducts	1	5
Schistosomiasis	1	13
Wilson's Disease	1	17
Associated with fibro cystic pancreas	1	13
Associated with endocrine disturbance	1	14

Of the cases in which no cause was found, 8 had portacaval anastomoses, and all have done well except the boy of 12 who died of liver failure five

years later The 4 patients with congenital fibrosis associated with proliferation of the bile ducts as described on p 21 did well after a portacaval anastomosis, in children with haematemesis from varices and a hard liver this condition should be suspected, and as it seems that the function of the liver does not deteriorate, the operation of portacaval anastomosis is strongly indicated, and should be done *before secondary thrombosis* occurs in the portal vein

anastomosis was performed After six months the spleen was no longer palpable and during the subsequent four years she has led the normal life of a *schoolgirl* *The histology of the liver is depicted in Fig 61*

In the cases of schistosomiasis and hepato lenticular degeneration, a portacaval anastomosis has prevented further haemorrhage for periods of *six and one year respectively since their operations The other 5 cases* were considered unfit for a venous anastomosis in view of the poor liver *function in 3 cases, and because there was thrombosis in the portal and* splenic veins in the other 2 Four of them are known to have died As it *is not always realised that patients with congenital absence of the bile* ducts can survive for a number of years, the following case is quoted

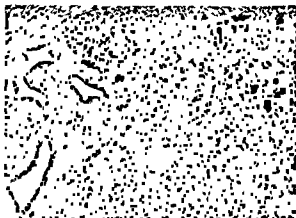
R L was admitted to hospital at the age of 6 weeks, jaundice having been noted at 3 weeks, but his stools had always been pale At a laparotomy the *gall bladder was shrunken and no common bile duct was found Over the*

bile ducts in the
...ch a case the liver
e hypertension

The 26 patients with *extra-hepatic obstruction have on the whole shown* disappointing results of treatment One patient who had portal pyaemia at the age of 15 started bleeding two years later, she has had a splenorenal anastomosis and an oesophageal transection and has remained well for one and a half years Another patient, a boy who has a stricture of the portal vein (Fig 8), had a splenorenal anastomosis five years ago and remains well A third had pneumococcal peritonitis at the age of 7 months, and probably at that time had portal vein thrombosis Two years later his spleen was much enlarged and he had his first haematemesis, bleeding continued at frequent intervals for the next two months and he died at the age of 2½ following an exploratory operation, his liver was



(a)



(b)

Fig 61 *Histology of the liver in a case of Congenital* Fibrosis

(*a*) Low power, showing many large bile ducts in the portal tracts and scarcity of portal veins × 30
(*b*) Higher power showing bile thrombi in the smaller bile ducts × 70

normal but the portal vein was represented by a spongy fibrous ligament although its branches in the liver were normal

The ages of the remaining 23 at the time of onset of haemorrhage are shown in the following table

AGE OF ONSET OF HAEMORRHAGE IN 26 PATIENTS UNDER THE AGE OF 20 WITH EXTRA HEPATIC OBSTRUCTION

Age of onset	*No of cases*
2	4
3	1
4	1
5	2
6	3
7	1
10	4
11	1
12	2
14	1
15	4
18	2

In none of these could any cause be traced but one child had had an acute suppurative arthritis of the hip during the first week of life and this may have been due to umbilical sepsis another child had acute osteo myelitis of the humerus when only a few months old and it is possible that his portal thrombosis occurred during that illness The portal vein in each case was replaced by a cavernous mass and in many the splenic vein was also thrombosed In 14 the spleen had been removed and in every case but one the bleed ng has recurred Fig 62 shows the fate of 18 of these patients who have died or who have been followed for more than five years from the time of their first haemorrhage It will be *noted that 7 of them are dead and the younger the age at first haemorrhage* the more serious the outlook Six patients who had splenorenal anasto moses when under the age of 10 have all had recurrence of bleeding

In this group nine venous anastomoses have been performed but the only ones which have so far proved satisfactory are four done in patients who had their first haemorrhages at 14 17 18 and 18 years respectively None of the splenorenal anastomoses in smaller children have remained patent Sixteen patients with extra hepatic obstruction who fall in this age group have had transection operations either of the oesophagus or stomach all these patients except one had had a previous splenectomy or splenorenal anastomosis and the bleeding had recurred From all these accounts it appears therefore that the outlook for a child with portal hypertension with extra hepatic obstruction is poor unless there is a suitable wide vein in the portal system to which an anastomosis can be

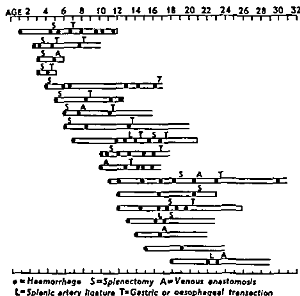


Fig 62 The fate of eighteen patients with extrahepatic obstruction who had their first haemorrhage before the age of twenty, and who have been followed for five years or more, or until death

made, this is most likely to be a splenorenal anastomosis If the splenic vein is small, the operation is not worth doing Makeshift anastomoses using other vessels have nearly always ended in failure Direct operations on the oesophagus or stomach offer the best hope in such patients

In children with intra hepatic obstruction the outlook, as in adults, depends on the function of the liver, but if there are large varices, or there has been bleeding and the liver function is reasonably good, an early portacaval anastomosis is strongly indicated before secondary thrombosis occurs in the portal vein Delay may mean that the only possible vein for an anastomosis becomes occluded, and the only opportunity of reducing the portal hypertension is lost

REFERENCES

JORDAN, P, PATTON, T B and BENSON, C D (1956) "Portal Hypertension in Infants and Children" *Arch Surg*, 72, 879–888

EKMAN, C-A (1957) "Portal Hypertension" *Acta Chir Scand*, Suppl, 222, 143 Stockholm

KOOP, C E and RODDY, S R (1958) "Colonic Replacement of Distal Oesophagus and Proximal Stomach in the Management of Bleeding Varices in Children" *Ann Surg*, 147, 17 25

SHUMAKER, H B and KING, H (1952) "Portal Hypertension in Children associated with Gastro-oesophageal Haemorrhage" *Arch Surg*, 65, 499–510

CHAPTER XII

RESULTS OF TREATMENT

It has been pointed out how difficult it is to judge the results of any form of treatment for portal hypertension, there are so many factors to be taken into consideration The question can be simplified to some extent if haemorrhage is regarded as the indication for operative treatment, for in that case freedom from further bleeding can be taken as the main indication of successful therapy Against this, however, must be set any other consequence of the operation the immediate mortality, any adverse effects on liver function, the incidence of neuropathy and the long-term survival This can be considered against the background that, without surgical treatment something over 50 per cent of patients with intra-hepatic obstruction will die within a year of their first haemorrhage, and Shumaker and King (1952) found that four out of five children with extra hepatic obstruction who had no operative treatment were dead within a few months In Chapters VII to IX many of the methods of treatment which have been advocated were described, it is unfortunate that in only one of these methods have reports of an adequate follow-up been reported, so this will be considered first

Portal-systemic anastomosis

The immediate mortality of this operation depends very largely on the selection of the cases, when the operation has been performed in the presence of poor liver function the risk is considerable Thus Blakemore (1952) found a post-operative mortality of 37 5 per cent in 16 patients whose serum albumin was below 3 gm per cent and 46 8 per cent in 32 patients who had ascites which had not responded to medical treatment This contrasts with an early mortality of 17 7 per cent in his first 135 shunt operations of all types when the serum albumin was above 3 gm per cent When the operation was performed for extra-hepatic obstruction the overall operative mortality was only 6 9 per cent compared with 21 9 per cent in the intra hepatic obstruction cases In his group the most common cause of death was liver failure, which seems to include portal-systemic neuropathy, the other major cause was operative haemorrhage which was more serious in the splenorenal anastomoses than in the portacaval operations Less common causes of post operative fatalities have been mesenteric thrombosis, and recurrent gastro-intestinal haemorrhage

In the writer's series of 111 shunt operations the most serious immediate complication has been portal systemic neuropathy, and the 4 cases of portacaval anastomosis who died from this cause have been described (Chapter 10) During the first three years that these operations were being undertaken (1947 to 1950) 2 patients died from operative haemorrhage following splenorenal anastomosis, but there have been no deaths from this cause amongst the last 100 operations Mesenteric thrombosis caused one death a month after a splenorenal anastomosis There have been no deaths directly attributable to the operation amongst the 85 cases of end-to side portacaval anastomosis apart from the 4 cases of neuropathy who died on the 7th, 16th 31st and 39th post-operative days respectively Operative mortalities recorded by other writers are 9 3 per cent in 75 cases (Ekman) 12 per cent in 141 cases (Linton and Ellis) and 15 per cent in 106 cases (Hunt) In the published series the operative mortality has been lower in the patients with extra-hepatic obstruction as these patients rarely have any defect of liver function, in consequence the overall mortality for splenorenal anastomosis is lower than that for portacaval anastomosis However, if these two operations are compared when employed in cases of intra hepatic obstruction it is found that portacaval anastomosis has the lower risk

Ekman has clearly demonstrated the vastly superior results of portacaval anastomosis when compared with splenorenal anastomosis when this aspect is considered In his follow-up from one to seven years there was a recurrence in 14 out of 32 patients with splenorenal shunts, compared with none following 20 operations of portacaval shunts Linton and Ellis, with a follow-up extending to nine years had 13 recurrences amongst 66 splenorenal shunts (19 5 per cent) and 3 amongst 26 portacaval shunts (11 5 per cent) Blakemore s figures were 6 after 52 splenorenal shunts (11 5 per cent) and 3 after 46 portacaval shunts (6 5 per cent) followed between one month and six years Our own experience is similar though the figures for splenorenal shunts are small Including the earliest cases where technique was imperfect and in some of which vitallium tubes were used, there are 19 splenorenal anastomoses who survived operation followed for periods up to ten years, 4 early cases were operated on for ascites and should be excluded, of the others 10 of these have had further haemorrhage but it should be borne in mind that 5 of these patients were children with small splenic veins and the view has already been expressed as a result of this experience that the operation is not worth doing in children As a contrast, amongst 81 patients with portacaval anastomoses who survived operation and have been followed from a few weeks up to seven and a half years only 4 instances of further haemorrhage have occurred One of these was fatal, and another patient died of cerebral thrombosis a few weeks after a haemorrhage, but the other 2 have remained well for five years following oesophageal transections

Three patients have died of unrelated disease and 4 others have died of liver failure

In a recent survey of all the patients who had had end to-side portacaval

piiot Some of the women have gone through successful pregnancies About 10 per cent have some ankle oedema and menstrual irregularities, these probably being a feature of their disturbed liver function In a few cases the liver disease is steadily progressive, particularly if the cause is not removed, those patients with haemochromatosis or Wilson's disease

Time since Operation	No of Cases	DEATHS 1st year	2nd year	3rd year	4th year	5th year	6th year	7th year	Survivors
Under 1 year	20	1							19
1 year	16	1							15
2 years	7								7
3 years	10	1	1	2					6
4 years	14	1		2					11
5 years	8	2							6
6 years	8	1			1				6
7 years	2								2
TOTAL	85	7	1	4	1				72

Fig 63 The survival of patients who have had end to side portacaval anastomoses

tend to run a slow downhill course, and the same happens to those alcoholics who do not mend their ways

Figure 63 shows the survival rate of 85 patients who had end to-side portacaval anastomoses, and Figure 64 gives the results of those patients who were operated on more than five years ago The appearance of the anastomosis in a patient who died three years after her operation is shown in Fig 65, and the venographic changes in another patient in Fig 66

Studies of liver function in most cases show no deterioration after the operation, the leucopenia and thrombocytopenia tend to persist but often in a less severe degree In all cases the spleen has become reduced in size and in about half it becomes impalpable two or three years after the operation

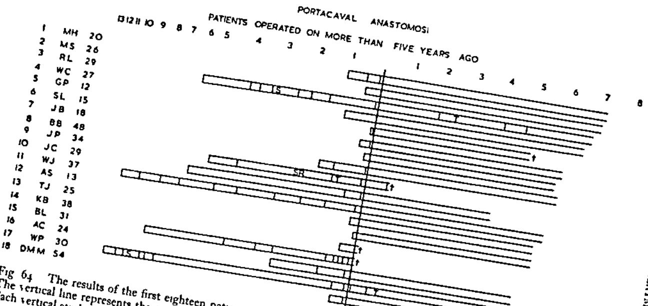


Fig 64 The results of the first eighteen patients treated by end-to-side portacaval anastomosis Five-year follow-up The vertical line represents the time of operation and the figures along the top the years before and after operation Each vertical stroke represents a haemorrhage The figures on the left give the age at the time of operation

Case 5 died of liver failure
Case 9 died of haemorrhage
Cases 14 and 15 died of neuropathy

S = Splenectomy
SR = Splenorenal anastomosis
T = Oesophageal transections

[Re]garding case 3, the last two haemorrhages have been slight and not required transfusion



Fig 65 The appearance of the portacaval anastomosis as seen from the caval side in a patient who died of a ruptured aortic aneurysm three years after her portacaval anastomosis

Transection operations and direct operations on the varices

Though the immediate mortality of these operations is low, whether carried out through the abdomen or thorax they are not nearly so efficient as a patent shunt in preventing further bleeding, the operation, however, carries considerable risk when performed as an emergency procedure in the bleeding patient Thus all 4 patients on whom the writer has carried out oesophageal transections as emergencies have died, but there has been no operative mortality amongst the 36 patients on whom it was an elective procedure Linton and Ellis (1956) however, saved 15 out of 20 patients on whom they carried out emergency suture of the varices The long term follow-up of these cases is also not encouraging though better than the expected survival if no operative procedure was undertaken In my own experience half the patients have had further bleeding but usually it is not so frequent or serious as before the operation, though

6 of the 36 survivors have died of haemorrhage between one and seven years after the transection operation Without quoting any figures, Tanner (1958) says that he has had excellent long term effects when gastric transection with freeing the vascular connection of the lower 5 cm of the oesophagus and upper 5 cm of both curvatures of the stomach has been carried out It looks as if this operation, which is more extensive than the oesophageal or gastric transections formerly carried out, will probably give better results, my own view is that the spleen should be removed at the same time if splenectomy has not already been performed

Hepatic artery ligation

This operation has been performed mainly in patients with rather poor liver function, where ascites has been the main symptom It might therefore be expected that neither the initial results nor the long term survival would be very encouraging, though in a few cases the relief of ascites has been dramatic

Altmeier and his colleagues (1955) in a series of 8 cases, most of whom had both haemorrhages and ascites, had an early mortality of 11 1 per cent, and 50 per cent of their patients were dead within three and a half years Results were considered good in only a third of the patients

Ruggieri (1955) had a post-operative mortality of 29 per cent in 24 patients, and half of his cases were dead within seven months, but the other half were regarded as clinical cures and returned to work

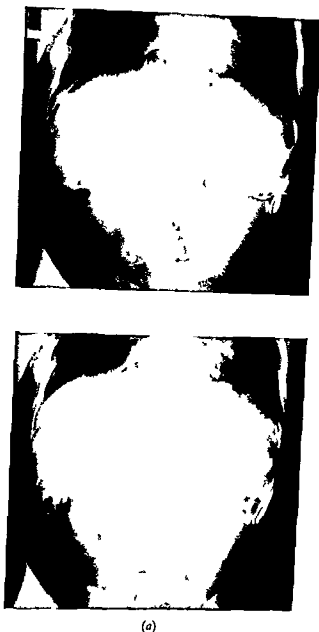
At the present time there is no means of determining beforehand which patients are likely to do well, and the results quoted may not be considered good enough to justify the high operative risk

Oesophago-gastric resection

Very few reports of the results of this operation are available, so it is difficult to draw any conclusion the immediate mortality has been rather high, and no five-year survival figures have been published In 8 cases reported by Macpherson, Owen and Innes (1956) there was no recurrence of bleeding, but the period they were followed was in no case longer than two years It has yet to be shown that the risks of the operation are justified by the long-term results and the disadvantages of the loss of the cardiac sphincter

Arterialisation of the portal vein

Attempts have been made to improve the blood supply to the liver by making a portacaval anastomosis and joining an artery, either the right renal or a graft from the aorta to the hepatic end of the portal vein This has been tried experimentally and in a few patients but the results have been uniformly disastrous and the operation has no place in the surgical treatment of portal hypertension



(a)

Fig 66 Trans splenic portal venograms (a) before and (b) after a portacaval anastomosis the time interval between the two plates is in each case 3 seconds





(*b*)

After the anastomosis the splenic vein clears more quickly and no opaque medium enters the collaterals

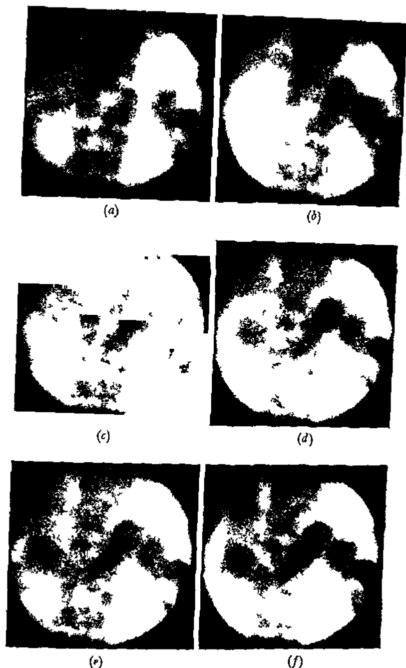


Fig 67 Shots from a ciné angiogram taken with an image intensifier Trans splenic venography in a case of intra hepatic obstruction ten



(*g*)



(*h*)



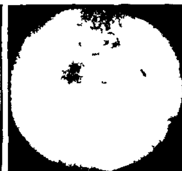
(*i*)



(*j*)



(*k*)



(*l*)

days after an end to side portacaval anastomosis (Same patient as Fig 39) In all shots from (*d*) onwards the vena cava can be seen but a little opaque medium still enters the tortuous left gastric vein

Injection of the varices

Most surgeons who have given this method of treatment a trial have abandoned it owing to the poor results and very few long-term successes have been reported in the literature

To sum up, we may conclude that the best treatment has been shown clearly by the results to be a wide portal systemic anastomosis using either the portal or the splenic veins These operations, unless carried out with meticulous care, are likely to fail owing to thrombosis in the neighbourhood of the anastomosis, and they should only be performed by surgeons with experience of the technique of vascular sutures The risk of occlusion is much greater than in the anastomosis of arteries This line of treatment however, does carry the risk of neuropathy which is occasionally quite disabling or may lead to lesser personality changes, and this risk has to be compared with the prospect of reasonable freedom from further haemorrhage If neither the portal nor the splenic vein is available because they are occluded, direct operations on the varices in the oesophagus or stomach or a gastric transection are worthwhile, but there is at present no one procedure which can be said to give results definitely better than the others A careful study and assessment of the long term results of these operations is needed during the coming years before the question can be solved In the meantime there is much scope for further study of the pathology of the liver, for in so many patients the prognosis depends on the functional capacity of this organ It is possible that steps to prevent umbilical sepsis may reduce to a limited extent the thrombosis of the portal vein which occurs in infancy and which leads to such tragic consequences as the children grow up

REFERENCES

ALTA [illegible] R C (1955) "The [illegible]

BLAK [illegible]
Surg Gynec Obstet, 94, 443–454

LINTON, R. R and ELLIS, D S (1956) 'Emergency and Definitive Treatment of Bleeding Oesophageal Varices" *J Amer med Ass*, 160, 1017–1023

[illegible] (1956) "Surgical Treatment [illegible] atment of Portal Hypertension in Liver Cirrhosis by Ligation of [illegible] Hepatic Artery" *Arch klin Chir*, 282, 1003–1013

TANNER N C (1958) 'Operative Management of Haematemesis and Melaena' *Ann R Coll Surg*, 22, 30–42

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